HYPEROSTOSIS FRONTALIS INTERNA: ITS RELATIONSHIP TO CEREBRAL ATROPHY.

By R. M. STEWART, M.D., F.R.C.P.Ed., D.P.M.,
Medical Superintendent, Leavesden Hospital.

[Received July 30, 1941.]

IN not a few of the recorded cases of hyperostosis frontalis interna there has been observed some degree of cortical atrophy, chiefly affecting the frontal poles, and its occurrence raises the question of its possible relationship to the osseous change.

Three alternatives may be entertained: (1) Brain atrophy is caused by the encroachment and pressure of the hyperostotic area on the surface of the brain; (2) brain atrophy is primary, the shrinkage of the frontal convolutions in some way exciting a compensatory bony overgrowth; (3) the two processes, osseous and neural, are entirely independent of each other.

There is little evidence in favour of the first view. While it is true that normally the apposition of calvarium to the surface of the brain is extremely intimate, so that the skilled anthropologist by study of the markings on the inner table can deduce important facts concerning the degree of development of the cerebral gyri, it does not follow that the hyperostotic zone must of necessity provoke pressure atrophy of the underlying frontal lobes. Unquestionably the process of bony accretion is very slow, and in its rate of growth can be compared with that of the hard fibroblastic tumour of the meninges. This tumour—the meningioma—over silent areas is capable of reaching a relatively large size without the production of obvious neurological signs, and it shows its effects on the brain not only by producing cortical atrophy, but also by an actual inward displacement of the affected area. It is difficult to believe that the slow-growing and widely distributed exostosis, if it were the cause of the frontal atrophy, would not also cause some degree of dislocation of brain substance. Moreover, reference to the literature of hyperostosis frontalis interna shows that, when present, the degree of cortical atrophy is never striking, and never in proportion to the amount of hyperostosis present.

For example, in the 71 cases presented by Dressler (1927) there was never more than slight atrophy of the frontal lobes. On the other hand, Hemphill and Stengel (1940) believe that pressure upon the brain surface contributes secondarily to the production of cortical degeneration. In the second of their

three cases reported in this Journal, they found " a moderate diffuse flattening in the frontal region corresponding with the projections on the inside of the frontal bone and a certain degree of gyral atrophy in this region." That this degree of atrophy must have been very slight is indicated by their failure to find microscopic evidence of any gross lesion in the cerebral cortex.

For the second view, namely that atrophy of the frontal lobes excites bony overgrowth, it has been pointed out that in many of the recorded cases the subjects have been persons of advanced age whose brains have shown some degree of senile atrophy, and that in conjunction with this atrophy their dorsal decubitus favours traction on the dura mater, and by creating a negative pressure excites the formation of new bone. This hypothesis has been well expressed by Greig (1928), who writes: " Why should the intracranial formation occur so invariably on the pars frontalis of the frontal? If the fullness of the brain plays any part in maintaining the approximation of the dura mater to the cranial surface—and that it must do so is apparent, since normally the skull is moulded to suit the brain—a senile shrinkage manifests itself anteriorly much more readily than posteriorly where the broader falx, the tentorium cerebelli and the great cerebral vein help to anchor it in position. The recumbent posture which in illness, weakness, or advancing years is more commonly and frequently assumed, would tend to relax the mechanical approximation of structures and promote an extradural serous effusion in which exuded cells might live, proliferate and function. In the anterior fossa of the skull the blood supply is more efficient than posteriorly, the venous channels of the diploë being supplemented by the meningeal vessels, the anastomosis from the face, the orbits and the nasal fossae. These factors seem to me sufficient to determine the site of the osteophytes. . . . The formation of intracranial osteophytes is an innocuous relief to a system supersaturated with calcium brought about in an area which age renders least important, and which has the necessary blood supply for the nourishment of bone-forming cells."

It must be observed, however, that Greig was concerned solely with museum specimens, nearly all of which were obtained from elderly persons, and that, consequently, in formulating a theory of the aetiology of hyperostosis frontalis interna he laid undue emphasis on the advanced age of its victims. When, as a routine measure, radiological examination is made of the skulls of living material, it becomes obvious that the condition is by no means confined to the elderly. In his series of 72 cases Sherwood Moore (1935) found an average age of 44 years. One of my patients was an epileptic boy aged 16 years. This is the earliest age at which the condition has been noted, but there are also on record other cases which have been discovered in early adult life, and consequently senility is not an essential part of the clinical picture.

If, as has been claimed, reduction of brain volume plays a part in the formation of the area of hyperostosis, it would be of considerable interest in an established case to know how the process would be affected by a superadded

gross atrophy *limited to one half of the brain*. The following case would appear to furnish the answer to this question:

Case report.—Annie H—, aged 68. Admitted November 22, 1932.

Following an attack of involutional melancholia in her fortieth year, the patient had, for 28 years, been an inmate of a mental hospital. When she came under observation at Leavesden she was an obese, hirsute, grey-haired dement of 5 ft. in height and 12 st. 12 lb. in weight. Physical examination revealed chronic bronchitis, myocardial degeneration and a marked degree of general arteriosclerosis. Blood-pressure 165/90. Blood Wassermann negative. The patient was childish in behaviour, confused and completely disorientated in time and place. Examination of her nervous system was negative, and she was regarded as an ordinary example of secondary dementia with no special features.

Progress.—On March 17, 1933, paresis of the left side of her face and slight weakness of the left hand were noted. Seven days later she presented a complete

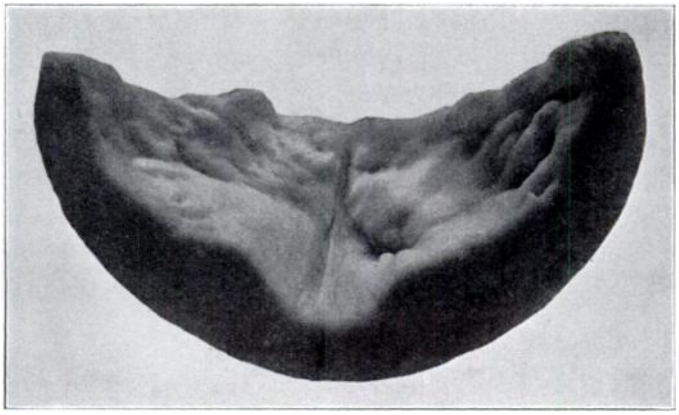

FIG. 1.—Part of the frontal bone, showing its extreme thickness and the growth of osteophytes on its inner surface. The photograph also conveys to some extent the more exuberant growth of bone on the left side.

left-sided hemiplegia. Lumbar puncture yielded a clear cerebro-spinal fluid, free from blood and under normal pressure.

The flaccid paralysis was accompanied by complete coma. Little restoration of function occurred in the paralysed limbs, and for the remaining three years of her life she was confined to bed, presenting a typical picture of residual hemiplegia. In the terminal phase of her illness dementia was profound and she was in every way helpless. She died on March 8, 1936—almost three years after the onset of hemiplegia.

Post-mortem examination.—Autopsy was held 22 hours after death.

The body was that of an elderly woman of obese proportions; the left arm and leg were in the typical hemiplegic attitude. The heart showed concentric enlargement of the left ventricle, fibroid degeneration of the myocardium and atheroma of the coronary arteries. There were calcified plaques in the carotids and an extreme degree of calcification in the peripheral arteries. The lungs showed hypostatic congestion, the liver an early nutmeg appearance and the kidneys an advanced stage of the arteriosclerotic type of chronic nephritis. The spleen was normal.

Skull.—The scalp and external surface of the calvarium were normal. Removal of the skull cap took considerably longer than normal owing to the density and

thickness of the bone. The strongly adherent dura mater was with difficulty separated from the anterior portion of the frontal bone. The freed calvarium was unusually heavy, 522 gm. in weight, and showed no vestige of sutures.

Frontal hyperostosis was present in an exaggerated form with an average thickness in the affected area of more than 2 cm. The osteophytic masses on the inner table presented a dense ivory-like appearance (Fig. 1), free from vascular channels, and extending as far back as the coronal suture. Lines of dural traction were present, and the medial groove of the sagittal sinus was not encroached upon.

Inspection of the two halves of the affected area showed that while a marked degree of symmetry existed, there was however a great difference in the degree of development of the osteophytic masses on the two sides, those on the left side being

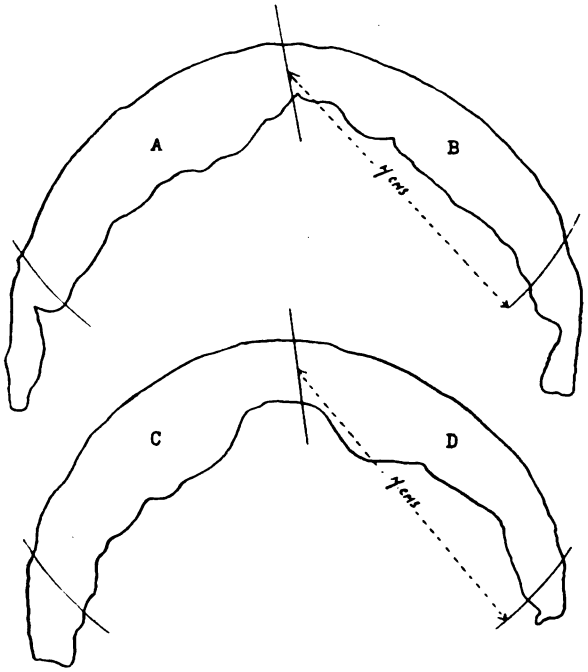

FIG. 2.—Drawings of two coronal sections of frontal bone from the area of hyperostosis, together with the measurement of each half expressed in square mm. Actual size.

more pronounced, so that on this side the average thickness of the bone was at least 2 mm. more than on the right. This difference between the two halves was better demonstrated when the bone was sawn into strips, the direction of the cuts being in the coronal plane and parallel to one another.

Fig. 2 shows outline drawings of two of these sections, together with their planimetric measurements in square centimetres. The two areas measured on each drawing were delimited by inscribing the arc of a circle with a radius of 7 cm. taken from a fixed point midway between the inner and outer tables, and in the medial plane above the narrow groove of the longitudinal sinus.

It will be seen that in each of the two drawings the area on the left side measured considerably more than on the right side, the difference being 1·68 cm. in the upper and 2.24 cm. in the lower drawing.

In addition to the frontal area of hyperostosis a few nodules were present on the inner aspect of the squamous portion of the temporal bone. The middle cranial

fossa likewise showed a number of sharply defined nodules (Fig. 3), these being more numerous on the left side. Slight thickening of the orbital plates was also noted.

The thickened dura mater was firmly adherent to the area of hyperostosis; elsewhere it was freed from the inner table without difficulty.

Brain.—On reflecting the dura, the brain was found to present gross shrinkage entirely confined to the right cerebral hemisphere, and occupying the territory supplied by the anterior cerebral artery and several branches of the middle cerebral (Fig. 4). The right frontal pole formed a shrunken mass in which no individual

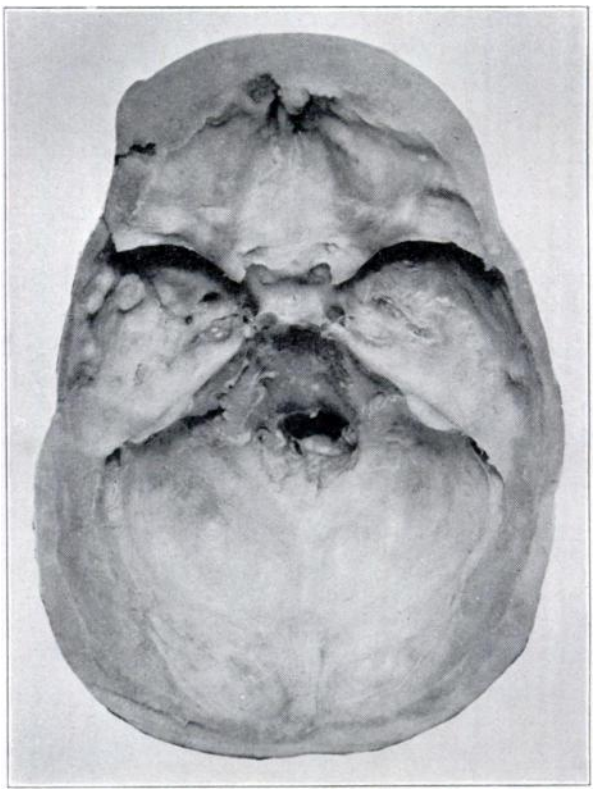

FIG. 3.—Base of skull showing nodular outgrowths in middle fossa.

gyri could be recognized and was covered by thickened pia arachnoid. On making a vertical coronal section through the hemisphere, it was found that the softening involved the basal ganglia and the wall of the third ventricle. Some idea of the extent of the atrophy is given by comparison of the weights of the two hemispheres: the weight of the right cerebral hemisphere was 253 gm., that of the left 465 gm.; total brain weight 840 gm. The left cerebral hemisphere showed a normal arrangement of gyri with slight generalized wasting of the fronto-parietal cortex.

At the base the large arteries showed marked arterio-sclerosis, the anterior cerebral vessels being especially affected.

The brain stem and cerebellum weighed 140 gm. Inspection of the medulla showed atrophy of the right pyramid.

Discussion.

In this case the post-mortem appearances left no room for doubt as to the pathological nature of the disease ; they were typical of an extremely advanced stage of hyperostosis frontalis interna in which the pars frontalis had reached a thickness of more than 2 cm.

In the living subject the thickness of the hyperostotic area can be measured on roentgenograms with only approximate accuracy, and repeated studies over a period of years appear to have been attempted by very few.

Lehoczky and Orbán (1938) made repeated radiographic examination of

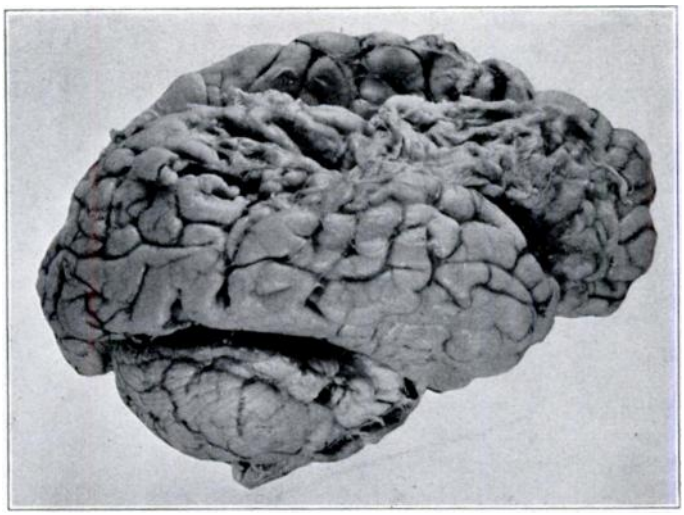

Fig. 4.—Lateral view of the brain showing massive softening of a large part of the right cerebral hemisphere.

the skull of a female patient suffering from this condition, and over a period of nine years noted the "slow progression of the bony changes."

Sherwood Moore (1936) makes the statement: "Neither progression nor diminution of the masses of bone has been observed in the few instances in which examinations have been made." The deposition of new bone on the inner table appears, therefore, to be an exceedingly slow process, and it seems likely that in the present case the excessive thickness attained must have been the product of a process in operation for many years. Some thickening on the temporal squama, on the orbital plates and in the middle fossa were present, but the feature of particular interest was the inequality of the frontal hyperostosis. This was quite obvious to the naked eye, and was confirmed by measurements made after division of the affected bone into strips.

If there is anything in the belief that brain atrophy excites a compensatory

deposit of new bone in this syndrome, it might be expected that a very severe degree of unilateral cortical atrophy occurring a few years before death would exert a far greater influence than the generalized and slight frontal wasting occasionally encountered, and consequently that the bony encroachment would be greater on the side of the gross atrophy, but as already described the appearances indicated that the thickening and nodular formation on the inner table was more marked on the side of the brain which had not undergone softening.

The question may perhaps be asked: Is it not possible that from the first and long before the date of onset of the paralytic stroke the deposit of new osseous tissue was more pronounced on one side than on the other, the difference being so marked that no subsequent added impetus to the deposit of new bone such as was provided by the massive softening in the cerebral hemisphere could possibly overcome this old-standing inequality?

For this view there is little support to be found in the literature, for practically all writers stress the remarkable symmetry of the process. It is true that when present nodular masses are not absolutely symmetrical on either side of the midline, but the two halves are almost invariably of equal thickness, such asymmetry as exists being the result of a slight difference in the extent of the two hyperostotic areas. It is, indeed, significant that very few writers have had anything to say regarding asymmetry. Greig is perhaps the only exception, for he states that he found the osteophytic growth more "distinct" in five out of 52 specimens in his collection, though evidently in none of them was the difference at all marked. In my series of 21 cases no other example of marked inequality was found.

If in the case recorded above the difference in the thickness of the two halves of the area of hyperostosis was not the outcome of unequal rates of osseous deposit extending over a long period of years, then it seems almost inescapable that in the three years subsequent to the onset of cerebral softening the osteophytic growth continued unchecked on the side of the calvarium which overlay the unaffected left cerebral hemisphere, while on the other side overlying the softened right frontal lobe the deposition of new bone was either brought to a standstill at the time of the cerebral thrombosis, or for the remaining years of the patient's life proceeded at a considerably slower rate. No certain opinion can be offered on these two alternatives, but the crossed relationship between the thicker half of the frontal bone and the shrunken half of the brain does seem to indicate that cortical atrophy plays no part in the causation of hyperostosis frontalis interna.

Summary.

In an advanced case of hyperostosis frontalis interna the thickening of the pars frontalis was very unequal, the osteophytic growth being much more evident on the left side than on the right.

The brain showed a massive lobar softening of three years' duration, limited to the right cerebral hemisphere, the other hemisphere showing only a minor degree of senile atrophy. The opinion is expressed that this crossed relationship between the gross atrophy of the right cerebral hemisphere and the thicker left half of the pars frontalis lends no support for the view that in hyperostosis frontalis interna, reduction of the volume of the brain stimulates the deposit of new bone on the inner table of the frontal bone.

REFERENCES.

DRESSLER, F. (1927), *Beitr. path. Arch.*, **78**, 332.
GREIG, D. M. (1928), *Edin. Med. Journ.*, **35**, 165.
HEMPHILL, R. E., and STENGEL, E. (1940), *Journ. Ment. Sci.*, **86**, 341.
DE LEHOCZKY and ORBAN (1938), *Brit. med. Journ.*, May 14, 1049.
SHERWOOD MOORE (1935), *Surg., Gynec. and Obstet.*, **61**, 345; (1936), *ibid.*, **35**, 30.
STEWART, R. M. (1928), *Journ. Neur. and Psychopath.*, **8**, 321.

https://doi.org/10.1192/bjp.87.369.600 Published online by Cambridge University Press