LOCALISED CRANIAL HYPEROSTOSIS IN THE INSANE.

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OF the various anomalies of texture which may be found in the cranial bones of the insane, generalised thickening and sclerosis is perhaps the commonest, and is seen in its most typical form in the dementias of epilepsy and chronic insanity. On the other hand, thickening of the cranial vault, limited to one particular area or bone, and accompanied in some cases by the formation of osteophytes, appears to be a rather rare condition. Usually it is the frontal bone which is affected in this way, and its precise appearance is subject to considerable variation. The construction of the calvaria may be fairly normal, the diploë being represented, but the two tables much thicker than normal. In other cases the increase is confined to the inner aspect, which has either a craggy embossed, or smooth surface. In a third type the main part of the enlargement consists of diploë limited by well defined tables. The amount of thickening is often considerable, and Shattock¹ mentions a case in which the maximum thickness of the frontal bone was 1.9 cm. According to Lawford Knaggs² in some cases the condition appears to be clearly inflammatory, the inner surface presenting a rugged, raised, irregular appearance suggestive of periosteal deposit, with channels directed towards the superior longitudinal sinus that evidently mark the position of dilated veins.

One of the earliest descriptions of localised hypertrophy is contained in Bevan Lewis' text book of mental diseases³, in which reference is made to fifty-four cases; in thirty-one of these the frontal was the site of the bony increase; in seven cases the occipital, and in six cases the parietal were the regions affected. Clouston⁴ has also recorded a particularly good example of hypertrophy limited to the frontal bone. The patient had suffered from alternating insanity of over twenty years' duration and the skull cap showed "enormous deposits of new osseous tissue distributed in an irregularly modulated way on the inner table of the skull." Concerning the origin of the condition Clouston writes: "This seems to be an aggravated example and type of what is almost universal in chronic insanity with periods of excitement. It is a proof of the structural effects of such repeated congestions of the branches of the carotid artery, even in the hardest tissue, and may be fairly considered to be of the same nature as the brain changes in the same cases, which are not so evident, but are no doubt far more important. The atrophy of the anterior lobes of the brain that usually accompanies such bony thickenings and deposits probably helps their growth, they being thus compensatory to some extent, like the increased cerebrospinal fluid."

Beadles⁵, in an interesting paper devoted to the morbid anatomy of the cranium, remarks that bony deposits when distinctly limited are usually found on the frontal bone. The condition, he states, is usually seen in cases of chronic mania or such as have passed into dementia, and frequently these patients have been the subjects of marked periodical excitement or alternating insanity. He agrees with Clouston that the condition is brought about by repeated temporary attacks of congestion of the brain and its coverings. A rather more adequate discussion of the pathology of frontal hypertrophy is given by Shattock, who describes in detail the appearances of a number of museum specimens. Among these there may be noted :—

1. The calvaria of an epileptic imbecile with a thickness of 1.9 cm. at the anterior part of the frontal bone.
2. A specimen with marked thickening of the inner tables of the frontal bone from "a stoutly built woman," age 37, who had epileptic fits every three or four days, and whose dementia was accompanied by attacks of violence.
3. A case of recurrent mania of fifteen years' duration, the patient dying at the age of 61. An irregular bony deposit was found on the inner portion of the frontal bone to the extent of three-quarters of an inch.
4. A female, insane for thirty-two years, who died in her 77th year. She is said to have been very stout. The calvaria was extremely thick, with little diploë; irregular projecting bony deposits were present on the inner surface of the frontal bone.
5. A female, age 55, with a history of alcoholism. Death occurred from dementia following mania of nine months' duration. The calvaria was thickened and congested, and the internal surface of the frontal bone very irregular from the presence of new bony growth.
6. A female, age 58, with a history of dementia following recurrent mania of over twenty years' duration. The calvaria was of average thickness, except at its frontal portion where the inner surface had a bossy or craggy surface.
7. A female dying at the age of 67 from senile dementia. Calvaria dense and heavy, with pronounced thickening on the inner aspect of the frontal bone.
8. A female, age 52. Chronic mania of seven years' duration. The calvaria showed slight general thickening, and a more pronounced hypertrophy in the frontal region with a craggy internal surface.
9. A female, age 72, who suffered from senile dementia. Thickening was present in the frontal region, which had a slightly irregular inner aspect.
10. Two specimens from Guy's Hospital Museum. One, from a female, age 55, with a long history of insanity, and the other, from a female, age 48, whose insanity was of eight years' duration. The brain was particularly small, and "marked with depressions corresponding with the elevations upon the inner aspect of the frontal region."

In the cases referred to above, little information is conveyed concerning either the physical state of the patients during life or the post-mortem findings, and it will therefore be of some interest to give a more detailed description of several cases I have had the opportunity of seeing during the course of the last fifteen years.

PERSONAL SERIES.

CASE 1. J. F., female, age 48, was admitted to an asylum on November 2, 1899, suffering from melancholia. There was a history of three previous attacks, the first of which had occurred at the age of twenty-one. No physical or neurological signs of importance were noted on admission, but her photograph showed a commencing lobulation of the skin on either side of the nose. At first she was fretful and querulous, and expressed a delusion that she was persecuted by a man. Her mental disorder ran an unfavourable course, and very soon she became dull, listless and apathetic, showing considerable impairment of memory. After two years a state of mild dementia supervened, and she was then reported to have become very ponderous and slow in her movements, and to have gained enormously in weight.

In November, 1911, I made the following notes on her condition: She is an extremely fat woman, weight 18 st. 4 lbs.; height 5 ft. 10 in. The distribution of adipose tissue is general, but the panniculus is particularly abundant on the abdomen, and there is a tendency to lobulation on the thighs. Her face has a swollen appearance, as striking as that of myxœdema, with large fatty swellings on its surface, giving rise to a marked narrowing of the palpebral apertures; there is no pitting on pressure. At first glance her bloated œdematous appearance suggests the presence of an intrathoracic neoplasm, but there is no cyanosis, her skin being pale in colour. Patches of yellowish pigmentation are present on the face and limbs, and in the former situation there are numerous small tags of loose skin, especially marked on the eyelids. The hair is scanty, harsh and dry, and is turning grey; on the upper lip and chin there are numerous coarse hairs, many of them being more than one inch in length; pubic hair normal in distribution and amount. No crescents are visible on her nails. There are no skeletal changes. Her voice is loud, harsh, and guttural. Appetite not excessive. Pulse 90, and of low tension. Heart sounds somewhat muffled; blood examination shows no abnormality. Lungs healthy. Temperature subnormal. Urine: sp. gr. 1010, pale in colour, no abnormal constituents; marked polyuria. Pupils equal, contracted, and reacting normally to light and on convergence. Special senses, cranial nerves, sensation and motor power normal. Reflexes normal.

Mental State.—Her general intelligence is difficult to estimate as she is suspicious and hostile in manner. When spoken to she uses obscene language, and pays no attention to questions. Although very torpid and sluggish she is able to do useful work of a simple nature, such as mending articles of clothing.

Subsequent History.—A noticeable feature during the last months of the patient's life was a marked polyuria. She passed large quantities of pale urine, free from albumen or casts. Owing to her incontinence the daily excretion could not be estimated. On November 6, 1913, she developed acute bronchitis, and on the following day her condition was one of extreme dyspnoea. At 9.30 p.m. she became unconscious and died on November 8, at 6.20 a.m. The left eye showed chemosis some hours previous to death.

Post-Mortem Appearances.—Necropsy was held five hours after death. The abdominal wall was several inches thick owing to the great deposit of fat, which was normal in colour. The calvaria was thickened, and exceedingly dense. The inner table was covered with large, irregular bony protuberances, most numerous on the frontal bone (Fig. 1), but also

Fig. 1. Case I. Showing bony masses on the inner table of the frontal bone.

present on parts of the parietals and occipital bone. The diploë was reduced in amount. The dura mater, which was slightly thickened and adherent to the inner surface of the skull, contained a large bony plate, nearly a quarter of an inch thick and more than one inch long, in the anterior end of the falx cerebri. Pia mater congested. The brain weighed 1,150 grm. and showed no naked-eye abnormality; the cerebral sinuses and veins were normal. The pituitary body was unusually firm and slightly enlarged. The heart showed an extreme degree of fatty infiltration, and the lungs evidence of acute bronchitis and oedema. The gallbladder was filled with cholesterolin stones. The ovaries appeared shrunken and cirrhotic; the suprarenals soft and friable. The kidneys and other organs showed no obvious departure from the normal.

Microscopic Examination of the Pituitary.—For the histological examination I am indebted to Professor T. P. Herring, who kindly sent me the following report:

“*Anterior Lobe*: There is marked sclerosis, a condition, in fact, resembling cirrhosis of the liver; there is marked deficiency in the chromophil elements and a strangulation of the cells by connective tissue.

“*Pars Intermedia*: The arrangement is rather unusual, and there is also a considerable spread of its cells into the pars nervosa. There is also a good deal of lymphoid tissue in it.

“*Pars Nervosa*: No abnormality save invasion by the pars intermedia.”

The right half of Fig. 2 illustrates the microscopic appearance of the anterior lobe; the left half shows for comparison the anterior lobe of a normal or nearly normal pituitary.

Fig 2. Right half shows sclerosis of anterior lobe of pituitary from Case I. Left half shows for comparison the cells of a normal anterior lobe. Both $\times 90$.

CASE 2. A. B., female, age 47, was admitted to hospital on May 17, 1911, suffering from "insanity with gross lesions" of six years' duration. She was extremely stout, her weight being 15 st. $3\frac{1}{2}$ lbs. Height 5 ft. $1\frac{1}{2}$ in. The notes on her physical condition are scanty. The presence of heart disease, and deformity of the fourth and fifth digits of the right hand are recorded, and reference is made to a stroke, said to have occurred seven years previously. Mentally she was in a state of dementia, being disorientated in time and place, faulty in her habits, and unable to occupy herself. In the ensuing fifteen years no change in her mental condition took place. Syncopal attacks occurred at infrequent intervals, and in 1921 the presence of right hemiplegia is recorded. This seems to be an error, there being no evidence of paralysis a year later. Her weight varied between twelve and fifteen stones.

On December 7, 1927, she became suddenly ill with distressed breathing, feeble rapid pulse, and pyrexia. Jaundice was present next day, and she died at 11.25 a.m. on December 9.

The autopsy was held on the morning of December 12, and owing to the cold weather the body was in a fairly good state of preservation.

The body was that of a fat female, with a heavy moustache and numerous coarse hairs on the chin. The distribution of fat was most marked on the lower part of the abdomen and thighs; girth at umbilical level $39\frac{1}{2}$ in. The skin had the greenish hue of jaundice. No deformity of the limbs was noted. The head measured $22\frac{1}{2}$ in. in circumference. The calvaria was thicker than normal, and this thickening was especially pronounced in the frontal region, being apparently due to overgrowth of cancellous tissue. Over a rather limited area of the frontal bone were irregular masses of dense bone. The sella turcica (Fig. 3) was much enlarged, chiefly at the expense of the posterior clinoid

Fig 3. Base of skull from Case II, showing enlargement of sella turcica.

processes, which together formed a thin plate which fractured easily. The pituitary gland was enlarged and fitted tightly in its fossa; its lateral aspects were compressed and flat. It was firmer than normal and weighed 0.875 grm.

The brain weighed 44 oz. Convolution pattern fairly complex; considerable wasting of the frontal and parietal convolutions, particularly in the left hemisphere. The right cerebellar hemisphere was markedly smaller than the left, and was associated with slight atrophy of the right olivary body. The arteries at the base showed a moderate degree of atheroma.

The lungs exhibited old pleural adhesions and scarring at the right apex; the heart weighed $6\frac{1}{2}$ oz., and showed fatty degeneration, and disease of the cusps of the mitral valve. The abdominal wall was heavily loaded with fat; the peritoneal cavity contained about 12 oz. of purulent material. A fistula existed between the apex of the gallbladder and the hepatic flexure of the colon. Facetted stones were present in the thickened gallbladder and in the common and cystic ducts. The liver weighed 62 oz., being moderately enlarged. On section the bile ducts appeared dilated and contained pus. The spleen and kidneys showed no noteworthy change. The ovaries could not be identified. Cause of death: general peritonitis following cholecystitis and cholangitis; fatty degeneration of the heart.

Microscopical Examination of the Pituitary.—The gland capsule is thickened, and lying on its external surface at the posterior pole of the organ is a collection of epithelium-like cells resembling those of the pars intermedia.

Anterior Lobe.—This appears considerably enlarged and shows a very marked increase in the amount of reticular connective tissue between the cells (Fig. 4). The latter

Fig. 4. Case II. Anterior lobe of pituitary, showing atrophy of cells and increase of connective tissue. $\times 90$.

are for the most part shrunken and reduced in number, there being a noticeable loss of the arrangement of cells in compact masses such as is seen in the normal gland. Granular oxyphil cells are fairly numerous, especially at the posterior end of the lobe.

Pars Intermedia.—There is less connective tissue in this area and in one situation the cells have spread into the *pars nervosa*.

Pars Nervosa.—This lobe is also somewhat enlarged, and the neuroglia more fibrous than normal. The interglandular cleft shows masses of colloid material.

CASE 3. C. K., widow, age 62, was admitted with a diagnosis of recurrent melancholia on June 2, 1909. Four years later she was noted to be childish, voluble and unable to remember recent events. A note made in February 1915 described her as more demented and incoherent. On December 9, 1919, she was noted to be walking with difficulty, and was unable to reply to questions or to obey commands. Slight paresis of the left arm and leg was present, and on January 1, 1920, her illness terminated with symptoms of uræmia.

A post-mortem examination was held eleven hours after death. The body was that of a very fat old woman, 11 st. 12 lbs. in weight. Height 61 inches. A considerable growth of hair was present on the upper lip and chin; the texture of the skin was smooth. The calvaria was dense and thickened, and on the inner table of the frontal bone there were dense nodular masses of bone. The brain weighed 1,160 grm. and apart from slight atrophy of the frontal gyri showed no abnormality. The heart and liver were fatty, the kidneys granular and contracted, and the lungs in a condition of hypostatic pneumonia. The pituitary gland appeared slightly enlarged; weight 0.56 grm.

Microscopic Examination. The microscopic appearances bore a close resemblance to those noted in Cases 1 and 2. The enlargement affected principally the anterior lobe; its cells were atrophied and surrounded by numerous strands of connective tissue (Fig. 5). The *pars intermedia* and *pars nervosa* showed no abnormality.

Fig. 5. Case III. Anterior lobe of pituitary showing appearance very similar to those in Case II.

In addition to the above personally observed cases I have had the opportunity of examining a number of museum specimens of frontal hypertrophy, and am able to give the following details of two examples. The clinical notes on the first case, the date of entrance into the asylum being 1875, were very meagre. The second was more adequately described.

CASE 4. F. S., male, age 19, was admitted on May 25, 1875, suffering from epileptic insanity. He was described as "A short, heavy-looking lad of good fatness." Mentally he was childish and peevish. His fits averaged two per week, and occurred only in the day time. He was employed in outdoor labour, until he finally became dangerous and partially demented. On July 9, 1902, he died in status epilepticus. At the post-mortem examination a tuberculous cavity was found in the right lung. The brain weighed 52 oz. and showed atrophy of the convolutions in the motor and frontal regions. Diffuse milkiness of the pia-arachnoid was also noted. The skull cap was exceedingly thick and dense, showing large deposits of bony tissue on the inner table of the frontal bone. These were distributed in an irregular fashion and were so hard that the chisel made little impression on them.

The cancellous tissue is principally responsible for the great thickness of the frontal bone (Fig. 6).

CASE 5. D. P., mechanic, age 51, was admitted to the asylum on August 3, 1885, suffering from melancholia; he had attempted to cut his throat. He was discharged after some months, but was soon re-admitted with a return of his suicidal tendencies and symptoms of melancholia, which became chronic. In 1892 he was described as having become very fat, and a further note made ten months later stated that he complained a great deal of frontal headaches and was now much stouter. Finally, he succumbed to capillary bronchitis on October 31, 1894.

Fig. 6. Skull cap from Case IV. Note hypertrophy of frontal bone; in this case the diploë is enlarged.

At the autopsy it was noted that he was extremely corpulent, measuring 53 inches round the abdomen; his height was 5 ft. 3 in. The heart weighed 18 oz. and was loaded with fat; the base of the aorta was atheromatous, and the aortic valve incompetent. The mucous membrane of the bronchi and bronchioles was acutely inflamed and the bronchial glands enlarged. The liver weighed 68½ oz. and was pale and fatty. The other organs showed no noteworthy change. The brain weighed 52½ oz. Slight dilatation of the ventricles was present. The pituitary body was not examined. The skull cap was extremely dense and thick, with irregular nodulated masses on the inner surface of the frontal bone (Fig. 7).

Fig 7. Case V. Cranial vault showing bony masses on inner table of frontal bone.

DISCUSSION.

As already noted, in the earlier papers devoted to this subject considerable importance is attached to the form of insanity displayed by patients who show hypertrophy of the frontal bone, and Beadles claims that the majority of, if not all, the cases occur in patients who have suffered from chronic mania or the dementia following mania. Analysis of the cases quoted above shows, however, that this is by no means invariably the rule, and other mental states such as melancholia or epilepsy may be present. For example, in case 1 of the author's series there was nothing in the history to suggest the "repeated attacks of congestion of the brain" said to exist in alternating insanity. Indeed, if the diagnosis can be trusted, in the early stages of her illness the patient suffered from recurrent melancholia, which according to Beadles is usually associated with thinning of the cranial vault. Even if the cerebral congestion accompanying mania occurred in every case, it would be difficult to explain how this could influence the supply of blood to the frontal bone, for the arterial supply of the calvaria is distinct from that of the brain, so that an active hyperæmia of the one does not necessarily involve that of the other. According to Shattock, the only way in which cerebral congestion could produce congestion of the calvaria would be by the engorgement of the cerebral veins which open into the various sinuses. It is, he thinks, conceivable that in this way a secondary congestion of the bones might, after the manner of Bier's method, bring about an overgrowth of their tissue.

A second factor supposed to be concerned in the production of localised calvarial thickening is recession of the subjacent cerebral substance accompanying the dementia seen in some of these cases, but how exactly atrophy or shrinkage of the brain can stimulate the formation of new bone is not easy to discern.

Atrophy of the cerebral convolutions is by no means uncommon in the chronic insane, but hypertrophy limited to the frontal bone is decidedly rare. Moreover, during the period of childhood or youth a cessation in the growth of any portion of the brain is not followed by thickening of the corresponding area of the skull. Again, shrinkage of the brain in adults is readily compensated for by an increased formation of cerebrospinal fluid, and the old assumption of a negative pressure within the cranium in cases of brain atrophy is hardly tenable to-day.

The connection between toxins circulating in the blood and the enlargement of certain bones, such as is seen in hypertrophic pulmonary arthropathy, has given rise to the suggestion that the condition under discussion may be toxic in origin. In certain cases a history of alcoholism is obtainable, and Shattock suggests that alcohol may serve as a second though subsidiary factor in producing congestion of the calvaria, or may act in the less direct manner of bringing a portal infection and a slow process of toxæmia, the congestion of a part with toxic blood being possibly more productive of overgrowth than one with normal. The principal objection to this view is the infrequency

of a history of toxæmia or alcoholism, and the occasional presence of frontal hypertrophy in mental defectives who have had no opportunity of acquiring the alcohol habit.

In the cases reported above stress has been laid on the presence of marked obesity. Possibly in some this can be attributed to the mental hospital regime, but in at least three the adiposity was of the pituitary type, being subsequently found to be associated with definite pathological changes in the pituitary body of an unusual type, and consequently the question arises whether the pathological and clinical features were causally related. The solution of this problem is by no means easy, for although nearly all observers are agreed that disease of the pituitary or of the neighbouring nervous structures at the base of the brain can effect profound alterations in the direction of overgrowth or undergrowth, the task of correlating the different types with pathological lesions is one of extreme complexity. In acromegaly, the prototype of hyperpituitarism, thickening of the calvarial bones is not a usual feature, although it is interesting to note that in the case of the acromegalic giant Charles O'Brien, whose skeleton is preserved in the museum of the Royal College of Surgeons, the skull shows marked thickening of the frontal bone. Nor does this condition appear to have been recorded either in Fröhlich's dystrophia adiposo-genitalis or in any of the other less well defined syndromes of hypopituitarism or dyspituitarism. The influence of the pituitary on the growth or rather the retarded growth of the cranial bones is, however, exemplified by the interesting syndrome of pituitary dysostosis, in which dwarfism, diabetes insipidus, and exophthalmos are associated with defective formation of certain bones of the skull. The condition occurs chiefly in young children, and is stated by Schüeller⁶ to be probably an unusual type of hypopituitarism.

If hypertrophy of the frontal bone could be shown to be a constant, or even frequent, feature in acromegaly it would be feasible to explain its presence in the insane on the assumption that overstimulation, resulting, among other things, in the formation of new bone, gave place to symptoms of exhaustion before sufficient time had elapsed for other bones to become involved, the symptoms of exhaustion manifesting themselves by the development of marked obesity. This does not, however, appear to be the case, frontal hyperostosis in acromegaly being the exception rather than the rule. None the less, the association of this condition with marked obesity in the insane—and especially those of the female sex—seems to be too frequent to be merely accidental, and when considered in the light of the pathological changes in the three cases recorded above, gives grounds for believing that we are here dealing with a hitherto undifferentiated type of dyspituitarism.

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