Morning Session, June 6.

HYPEROSTOSIS CRANII (OPHALOMEGALY),
WITH ILLUSTRATIONS.

BY DR. JAMES J. PUTNAM, OF BOSTON.

ABSTRACT.

He reported four cases of this disease, which, to judge from published records, seems to be a rare one in its typical form, though instances of partial hyperostosis and exostosis are not very common. The most important literature on the subject consists in an analysis by Virchow in his treatise upon tumors; a monograph by Baumgarten on Leontiasis Ossea, published in Paris, 1892; a recent paper by Dr. M. A. Starr in the *American Journal of Medical Sciences*, for December, 1894.

The physiological characteristics consist, primarily, in a thickening of the bones of the head which usually occurs for the most part outwardly, though there is always more or less unevenness of the inner surface and sometimes considerable encroachment on the cranial cavity. In addition to this, an interstitial growth of the bone commonly takes place, destroying the diploe and forming a dense homogeneous tissue. In consequence of these changes the shape of the head becomes greatly enlarged, especially anteriorly and laterally. The superficial cavities and apertures of the skull are, as a rule, more or less completely filled up, so that the eyes are pushed outward and the nerves and vessels entering and leaving the cranium are compressed. The disease is chronic in its duration, beginning frequently, though not variably, in youth, and lasting often twenty or thirty years.

There are several types of the bony enlargement, of which the chief are the leonine, in which the prominences of the upper face become large and rounded, and that in which the thickening is confined to the vault of the head.

Of the cases described by the writer all belong to the latter category, except one, in which the malar

bones were prominent, but in two of the cases the orbits were more or less encroached upon.

The disease seems sometimes to originate in an inflammatory process, following injury or disease of the soft parts. Often, however, no special cause can be determined, and Virchow, while considering the affection in a general sense as inflammatory, refers to the fact that in some persons a predisposition to it seems to exist.

In Dr. Starr's case, besides the changes in the bone, there were alterations in the soft parts, such as an increase in the size of the neck due in part to enlargement of the vertebræ, and nutritive disorders of the skin, which suggested to him the possibility that the disease might be a general dyscrasia, analogous in a distant sense to acromegaly or myxœdema. Baumgarten had advanced a similar idea. In the writer's cases no distinct changes of this sort were present. Occasionally other bones of the skeleton, and, indeed, the whole osseous system show a similar tendency to overgrowth. Histologically, however, this is not to be distinguished from an inflammation, and differs somewhat from acromegaly.

The symptoms of the affection are mainly the following: Exophthalmus, deafness, blindness, facial paralysis, headache, double optic neuritis, suppuration in the ears or elsewhere, epilepsy, the attacks sometimes occurring very early, vertigo, a peculiar giving way of the leg, disturbance of respiration and mastication; loss of teeth; drowsiness; mental deterioration. Not all these symptoms are, however, present in each case.

The writer showed the photographs of three of the cases described by him, a specimen of the cranium of the fourth, and one patient was demonstrated before this Society. A case with autopsy, communicated to the writer by Dr. Morton Prince, was also read, and a remarkable skull showing the peculiarities of the disease, preserved in the Warren Museum of the Harvard Medical School, was also shown and described.

CASE I.—Female patient. No history of syphilis or other constitutional disease, except that when three years old she had caries of the ankle. It is possible that her mother and sister died from the same disease with the patient. The first positive symptom was noted when the patient was nineteen years old, though even before this she had suffered from excessive drowsiness. The

following symptoms appeared gradually and in the sequence indicated: Headache, loss of teeth, loss of hearing, discoloration of the skin. From near the beginning of the case there had been a sero-purulent discharge from both ears, and at the same time the head had begun to grow broader and the eyes more prominent. When examined by the writer, nine years after the affection began, double facial paralysis was present, and cataract of one lens. The gait was feeble and uncertain, but the hands were used freely. No sensory symptoms were present. The patient grew gradually worse, and finally drifted into the Cambridge Almshouse, where she died. For some months before her death she had suffered from chronic suppurative diffuse inflammation of the pericranium. Cerebral symptoms have never formed a marked feature of her case, and double optic neuritis was never present, so far as is known, certainly not when she was first examined. Through the kindness of Drs. Bryant and Holt the cranium was obtained for the Warren Museum. It was found to be relatively globular in shape, owing to broadening in the parietal region, and everywhere the walls were thickened, but much more anterioral than elsewhere. The forehead was prominent, and the malar processes enlarged; the orbits narrowed and shallow. The surface was everywhere uneven, but otherwise smooth. Almost all signs of sutures had disappeared. Many of the cranial apertures were narrowed or destroyed. The size of the cranial cavity was not greatly diminished, but the bony prominences were almost universally thickened and the angles rounded off. The sella tursica was smaller than normal and altered in shape. The surface of the frontal bone and the lower half of the temporals was worm eaten by caries, due to the suppurative process above alluded to, and the same process had nearly destroyed both the zygomas. The vascular grooves were practically absent, and the diploe converted into a dense homogeneous mass of bone.

CASE II. is of special interest from the fact that the disease began in infancy, the two exostoses which afterwards became so prominent, having been noticed when the patient was five years old. It is also noteworthy from the fact (alluded to also by Baumgarten) that he began to suffer from convulsions in his seventh year, before the bony growth had attracted attention. These convulsions did not recur with such frequency as to sug-

gest an ordinary case of epilepsy, though toward the end of his life, when he was about twenty years old and his disease far advanced, they became somewhat more frequent. The other symptoms consisted in dull headaches, impairment and finally entire loss of hearing, double optic neuritis of an excessively chronic course, ending after many years in blindness. Toward the end of his life he became dull and irritable. The gait was at first uncertain and then progressively feebler, so that he was finally confined to his chair. There was slight prominence of the eyes. The hyperostosis was diffuse, but was also manifested by the appearance of two large exostoses in the neighborhood of the vertex, which increased steadily in size. He died when about twenty-two years old. No autopsy was allowed.

CASE III. is that of a man, still living, who was shown at the meeting. He is fifty-three years old and gave the history that his head has been growing larger for about fifteen years. The principal symptoms have been pain, nausea and vomiting, and poor hearing. He has also been greatly troubled by a throbbing sensation in the right ear which is almost constant, unless he compresses the carotid, which relieves it. He wears a hat which is more than an inch larger than formerly, and so large that it has to be made to order. The forehead is very prominent and usually covered with moisture, and is marked by large veins, which swell up on the least vascular excitement or from slight compression of the neck. The eyes are not particularly prominent. The surface of the head is uneven from small exostoses and the thickening seems to be universal.

CASE IV. is that of a woman, thirty-five years old. The enlargement of the head was first noticed twelve years ago or more, and is now greater than in any of the other cases, though the symptoms of discomfort are less marked. In fact, the only symptom of note has been a tendency to severe burning sensations on the right side of the head and face, with a sense of numbness in the face, recurring in attacks of a few moments duration. Besides this she has a feeling of debility and a sense of fullness in the head. The enlargement is universal, but, as in the last case, it is of the cranial type, not affecting the face below the forehead, and not causing noticeable prominence of the eyes, though the edge of the orbits are thickened and rounded. The surface of the head is very irregular and is covered with bony

prominences of different shapes. There is a thick growth of coarse gray hair, but otherwise the nutrition of the skin and its appendages seems unchanged. In none of these cases was anything observed analogous to the general trophic changes seen in Dr. Starr's patient, except that in the first case the skin was of a yellowish color; in the second, that the neck was rather thick, and in the third, the hair coarse.

DISCUSSION.

Dr. STARR, of New York.—I have nothing to say, Mr. President, excepting that these cases are more advanced and more extreme than the case I reported in the *JOURNAL* to which I believe Dr. Putnam has referred. The heads are much larger. I think that the pathology of it is perfectly well shown by these sections of the skull. The origin of it I do not suppose any of us know anything about. And as for treatment I don't suppose any one knows any more about that. I tried thyroid extract with my patient because I had some rather remarkable and interesting results in acromegaly by giving thyroid purely as a tentative measure, but in my patient the thyroid treatment did not do any good whatever. My patient died of pneumonia, and I did not secure an autopsy.

Dr. PRINCE, of Boston.—Apropos of the last remark of Dr. Putnam I will briefly narrate a case, an account of which I have already written for Dr. Putnam, but which I have not with me. In this case the enlargement was not as general as in Dr. Putnam's cases. About three years before I saw this patient he met with a severe accident, viz., he was hit on the forehead by some tackle, and the story was that he had not been quite himself since then. At the time I saw him he suffered from symptoms of brain pressure very similar to those of cerebral tumor,—nausea, vomiting, headache, somnolence at times, loss of memory, and more or less of dementia. I saw him in consultation with a view to operation. In his case there was a large protuberance largely confined to the frontal bone. It was evident that the parietal bones were enlarged. Afterwards I was sorry that I decided against operation. I did not know how extensive the *ostosis* might be, and it seemed as if the operation would be a difficult one. Afterwards he died. There was no paralysis of any kind when I saw

him, but at the time of his death I was told by his wife that besides a certain amount of dementia and loss of memory, paresis of the left side of the body had been present. When we made the autopsy we found the thickening was almost entirely in the frontal bone. The orbital plates were enormously enlarged, so that they sprang up well into the cavity of the skull and left a very small space between them. The brain was squeezed downwards into this space between the orbital plates. The convolutions were flattened and smaller than normal. It was evident that the brain was compressed, pressed upward and downward. I thought from the examination of the skull that it would have been feasible to have relieved the condition by operation, and in such cases as this I should say an operation could be performed. That certainly was the impression of all who saw the autopsy, though the case was probably an exceptional one. There was no history of syphilis in this case, if I remember rightly. There was marked exophthalmos, but the man said his eyes always had been prominent.

Dr. HAMMOND, of New York.—I should like to ask if Dr. Putnam has seen in his cases evidences of structural changes in the skeletal apparatus elsewhere in the body. In a case I have seen recently almost the counterpart of the one presented here, there were five or six of these enlargements, and the skull was fissured and cracked in all directions. He was a syphilitic case, and four years previously, while driving, his horse stumbled,—he was driving with one hand,—he jerked the reins and broke his humerus; and about a year after that in pulling a cork out of a bottle, he broke his clavicle. The bones showed wherever they could be felt, enlargements and nodosities, and it is quite probable that in many of these cases the same cause that produces the structural changes in the bones of the skull does it elsewhere in the skeleton.

Dr. PUTNAM, of Boston.—That is one of the interesting points in connection with this matter, and I presume where syphilis is connected with the case the latter should be in a different class from those where no such cause is present. In a certain proportion of cases there does seem to be a tendency to enlargement of the bones through the rest of the body and changes of nutrition of various sorts. Virchow alludes to that fact, although the case on which he relied has been made out to be a

case of acromegaly probably. In Dr. Starr's case enlargement of the vertebræ was noticed. There is a tendency to bony enlargement. That would seem to militate against the view that the disease is distinctly due to spread of inflammation from some special point of injury. Possibly some toxic process goes on which affects the osseous system at large. Virchow, although he thinks the disease is inflammatory, speaks of predisposition in certain persons, and he means predisposition to include the tendencies related to certain epochs of development and other influences of that order, and that would include a good deal of what we now mean under the head of dystrophic tendency.

Dr. MILES, of Baltimore, showed

A CAST OF A BRAIN AND A BRAIN PRESERVED IN ALCOHOL.

Dr. WORCESTER, of Danvers, Mass.—During my service at the Arkansas State Lunatic Asylum I had the opportunity of making autopsies on three patients, in whom there was a very decided asymetry in the two hemispheres. In one case one hemisphere weighed seventeen ounces and the other nine. I will not say there was asymetry of the cerebellum in those cases. I examined the cerebellum in each of those cases and there was nothing that attracted my attention as to a difference in size between the different hemispheres of the cerebellum, although I might have overlooked a slight difference, I do not think I should have been likely to overlook any very striking one.

Dr. MILLS, of Philadelphia.—Many years ago I exhibited to the Philadelphia Pathological Society the brain of a negro murderer who had been confined in prison in that city for some time. In this case one of the most marked peculiarities of the brain was the atrophy of one cerebellar hemisphere co-incident with atrophy of the opposite cerebrum and particularly in the motor region. In a limited area of the pons was an old hæmorrhagic cyst, apparently the only active lesion which had been present in the case in this patient, and it seemed to me that this cyst in this particular locality had determined the atrophy both of the cerebellum and of the cerebrum.

Dr. STARR, of New York.—I regret that Dr. Peterson is not here to add to this discussion. Dr. Peterson has a

very large service at Randall's Island in the Children's Hospital for Idiots and has a number of these brains. I am indebted to him for photographs which I show to my students, of atrophy in various parts of the brain, and they are usually, I think, uniform in this, that you find an atrophy of the cerebellum corresponding to an atrophy of the opposite cerebral hemisphere, and you may depend upon it that the condition is congenital or acquired before the fifth year. Secondly, Dr. Peterson has some beautiful specimens of atrophies in different localities, and it is interesting to find that in these congenital cases the atrophy of the cerebellum accompanies only atrophy of the frontal or motor zone. If there is atrophy of the temporal or occipital region congenitally, we have no atrophy of the opposite half of the cerebellum.

Dr. BOOTH, of New York.—I have a case in point which I reported two years ago. In the brain an acquired cyst was developed after infection from scarlet fever at the age of five, and the post-mortem was held at about the age of thirty-three. There was a diminution in the left cerebral hemisphere and in the right cerebellar hemisphere. And the diminution either in the cerebral or cerebellar hemispheres was in the same proportion, not simply difference in weight, but the proportion of loss was the same.