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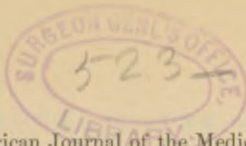
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## MEGALO-CEPHALIE, OR LEONTIASIS OSSEA.

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WITHIN the past few years attention has been drawn to various disturbances of nutrition occurring in the human body, either limited to certain parts or affecting the entire organism. We are at present familiar with changes in the growth and size of single parts or segments of the limbs, such as are seen in the various forms of atrophy and hypertrophy of muscles or of the skin, and also with changes in the appearance and size of the hands and feet, such as appear in acromegaly. Disturbances of nutrition of a more diffuse kind, attended by changes in the size and appearance of many tissues, are known to occur in myxœdema; and in fact, it is the study of this latter disease that has thrown much light upon the nature and origin of all disturbances of growth and nutrition. The fact that the secretion of the thyroid gland, when absorbed into blood, has a direct influence upon the nutrition of the hair, the skin, the subcutaneous tissues, the muscles, and many of the internal organs, cannot be denied. And there are many facts which show that an excess of thyroid activity is just as detrimental to the human organism as an arrest of its functions, for it seems quite apparent that in certain of the cases of Graves' disease some of the symptoms of malnutrition are to be ascribed to an increase in thyroid activity. The facts ascertained regarding myxœdema and its dependence upon the thyroid gland lend a certain amount of probability to the statement that acromegaly is dependent upon disease of the pituitary body, although this is not fully proven. We do know that the progressive growth in size in the hands, feet, and bones of the face, is often found associated with a hypertrophy of the pituitary body. Again, it is probable that all the symptoms of Addison's disease are definitely to be assigned to disease of the adrenal bodies. We are warranted in the surmise, in view of these facts, that there are certain diseases of nutrition which are dependent upon lesions in various glandular organs, whose function has been hitherto obscure, which deserve further study.

There has lately come under my observation, in association with Dr. G. K. Dickinson, of Jersey City, a case of a peculiar character, which, so far as I know, has not as yet been described, of a trophic disease quite analogous to acromegaly, the chief symptom of which is a gradual or

progressive enlargement of the head and neck, and which I venture to name megalo-cephalie.

FIG 1.



Condition in 1889, before enlargement of head and neck began.

The patient is a woman, aged fifty-two years, American by birth, and of healthy parentage, her mother being still alive at the age of seventy-

FIG. 2.



Present condition, 1894, showing uniform enlargement of the head.

eight. She has one brother and two sisters living and healthy; the family history is free from all evidence of cachexia or neurotic habit;

she has been married since the age of twenty-one, but has had no children; she has been in excellent health until the beginning of this disease—six years ago. The first symptoms noticed were formication felt in the tips of the fingers, particularly of the right hand, which gradually extended upward to the shoulder; this numbness was associated with a slight clumsiness of movement and uncertainty in tactile perception, although there has never been and is not now any actual anæsthesia, either of touch, temperature, or of pain, and the muscular sense is normal. She has never felt any of this numbness in the legs or body, but she has noticed for the past five years a disturbance in the gait, consisting of an uncertainty in movement and a subjective feeling of weakness. During all this time there has been a slowly progressive enlargement of the head and neck, and at the same time a gradual diminution in stature. Five years ago she was five feet seven inches in height, now she is five feet

FIG. 3.



Present condition, 1894, showing enlargement of the neck and head.

two; this shortening is partly due to a stooping posture resembling that assumed in paralysis agitans, partly due to an apparent sinking of the head and neck between the shoulders, which causes a very peculiar appearance.

The striking feature of this disease and its chief characteristic, as shown in our patient, for whose history I am indebted to Dr. Dickinson, is a slowly progressing increase in the size of the head, face, and neck, both the hard and soft tissues being apparently affected. The skin is diffusely thickened to a slight extent, is thrown into folds and not merely stretched over the enlarged bones, as it would be were it not involved in the process; and while it is not rough or scaly, is slightly pigmented and has something the appearance of the skin in myxœdema. The sub-

cutaneous tissue is also diffusely thickened, so that it throws the skin out and forms a loose body beneath it. The bone appears to be the portion primarily and chiefly affected, just as in acromegaly. There is apparently a general hypertrophy of the bone over the entire calvarium, more marked just behind the temples. This is not limited to definite small areas, but is quite uniform everywhere, so that the skull is enlarged uniformly. The head measures twenty-four inches in circumference about the forehead, fifteen inches from ear to ear over the vertex, and sixteen inches from the root of the nose to the occipital protuberance. Even the vertebræ of the cervical region appear to take part in the enlargement, and as a result the head is less movable upon the spine than it should be, and the spines of the vertebræ from the fourth to the seventh are decidedly prominent in the neck. A feeling of weight in the head is quite intense and had led to the hair being cut short, and it was noticed that the hair had become quite thin, especially at the sides, when formerly it had been thick and heavy. This diffuse growth of the head and neck is quite well shown in the photographs, which are all the more instructive, as the contrast between the woman in her ordinary state of previous health five years ago and in her present condition is shown. The thickening of the facial tissues tends to decrease the natural mobility of the face, so that the facial expression is more fixed than in a state of health. The thickening seems to have encroached upon the cavities of the nose and mouth, there being some obstruction to breathing and swallowing without thickening of the tongue. There is a hard respiratory sound in the trachea constantly in breathing, and this has been present for a year past.

The general symptoms which this patient suffers from are gradually increasing mental irritability and general nervousness, without any mental or emotional defect; an increasing disability to walk and to use her hands, attended by much numbness, especially in the hands, an occasional pain in the neck, arms, and hands; some awkwardness of movement in the fingers, but no marked ataxia, for she can knit, can thread a needle and do fancy work. There is no anæsthesia present, and there is no loss of pain or thermal sense. The act of walking is performed with difficulty and is now only possible with the aid of holding a chair, though there is no ataxia of movement and no spastic rigidity of the legs; the knee-jerks are slightly increased, but there is no clonus; the control of the sphincters is perfect. In walking she has a general sense of insecurity and feebleness, but there is no evidence of paralysis. She stoops forward much like a patient suffering from paralysis agitans, but has no tendency to fall or to festination. She is able to walk up and down stairs with help. Her neck is sixteen inches in circumference—at least four inches larger than formerly. There are two soft non-fluctuating swellings below the ears having the feel of dif-

fuse, fatty infiltrations, and the neck is uniformly increased in size. There is no apparent swelling of the thyroid gland. The cervical vertebræ feel thick and are unduly prominent, but are not tender. She has had considerable pain along the course of the occipital nerves; in other respects her general health is very good, the digestive and urinary functions being perfect. The heart is regular, is not enlarged, and the pulse tension is normal. There are no cardiac murmurs, but a blowing murmur is heard over the arch of the aorta synchronous with the first sound. There is no area of dulness, and there are no other symptoms to suggest aneurism. No cause is to be found for this condition, which has advanced so gradually that its actual date of onset is difficult to determine, though she is certain that it began more than five years ago.

The very extraordinary appearance of the head in this patient suggested to me that she might be suffering from a condition known as leontiasis; but a careful study of the literature regarding leontiasis convinced me that under this name a number of entirely different conditions have been grouped.

The term leontiasis has been applied to a form of leprosy affecting the face, and very clearly described by Leloir in his monograph upon lepra, where several plates illustrating the appearance of the face in this disease are to be found. In this condition there occurs a deposit of new material here and there upon the face beneath the skin, and these deposits gradually enlarge and coalesce until the entire face is changed into a hard mass with deep seams and roughened surface, the features being almost obliterated and the eyes sometimes closed. The bones of the face are not usually involved, and the disease does not extend upward beyond the line of the hair, and does not produce any enlargement of the cranial bones. The term leontiasis has been applied to this because the rough, seamed, and thickened appearance about the mouth suggests the idea of the jaw of a lion. It is said that the term has been in use since the time of Galen.

There is a second condition to which the term leontiasis has been applied—namely, a condition of elephantiasis affecting the tissues of the head and neck. This is mentioned by Virchow in his work upon tumors. He describes it as a diffuse thickening of the soft parts of the head and face, consisting of an increase in fibrous tissue, and considers it a form of fibroma molluscum. This form corresponds in its history to elephantiasis; it begins quite suddenly with an acute condition, which may be either erysipelas or dermatitis or phlebitis; the skin becomes swollen and red simultaneously, and there is often an eruption of vesicles after the acute symptoms subside, but the swelling does not disappear entirely, and the skin pits upon pressure. Later, there is a recurrence of the acute condition and an increased swelling; and so the

disease goes on with acute exacerbations and remissions; ulceration of the surface and very deep pigmentation are commonly observed in the parts affected. After describing this condition and recording a clinical case described by Gruber, Virchow says: "I do not wish to affirm that there is not a form of leprosy which calls to mind the appearance of leontiasis; on the contrary, I have often seen such a form of leprosy in Norway, but the greatest deformity occurring ordinarily without ulceration is the peculiar characteristic of fibroma molluscum, or elephantiasis molluscum."<sup>1</sup>

I think it is quite evident that the disease from which our patient is suffering does not in any way correspond to either of these two diseases to which the term leontiasis had been applied.

There is a third disease to which Virchow gives the name bony leontiasis (*leontiasis ossea*), which, however, resembles *megalo-cephalie* more closely. In his description of tumors of a bony nature, after describing exostosis and periosteal thickenings, Virchow devotes a section to hyperostosis of the face and head. This may take two forms: first, a diffuse hyperostosis of the cranium; secondly, localized hyperostosis upon the maxillary bones. This latter condition we may dismiss without further consideration, inasmuch as it presents no points of a resemblance to our own case. It has been studied carefully by Le Dentu in *La Revue mensuelle de Médecine et de Chirurgie*, 1879. The diffuse hyperostosis of the cranium is, according to Virchow, symmetrical and uniform, so that it does not present any bony excrescences and suggests the idea of tumor. Virchow's description is based upon the examination of certain skulls, which are preserved in various museums, the first one having been described by Malpighi, 1697. There are about six of these skulls known in Europe, all of them presenting tremendous enlargement of the bones of the head of a diffuse character. Virchow says: "This condition is for the skeleton what leontiasis is for the soft parts; the appearance of these pathological specimens, seen a certain distance, produces the effect of leontiasis modelled in plaster. Hence, I have the less hesitation in giving the name of bony leontiasis, since, as I believe, the disease corresponds exactly with that which we call elephantiasis in the soft parts. Ordinarily, the hyperostosis is associated with a fibrous change in the soft parts; in the rare cases which we have seen, it is the periosteum which is the principal seat of the disease. Unfortunately, we have no records of clinical observations, and no knowledge of the exact conditions present by recent autopsy."<sup>2</sup>

In a monograph upon *leontiasis ossea*,<sup>3</sup> Baumgarten has made a

<sup>1</sup> Virchow: *Pathol. des Tumeurs*, vol. i. p. 324.

<sup>2</sup> Virchow: *loc. cit.*, vol. ii. p. 22.

<sup>3</sup> *La Leontiasis Ossea*, par Dr. F. Baumgarten, Paris. Steinheil, 1892.

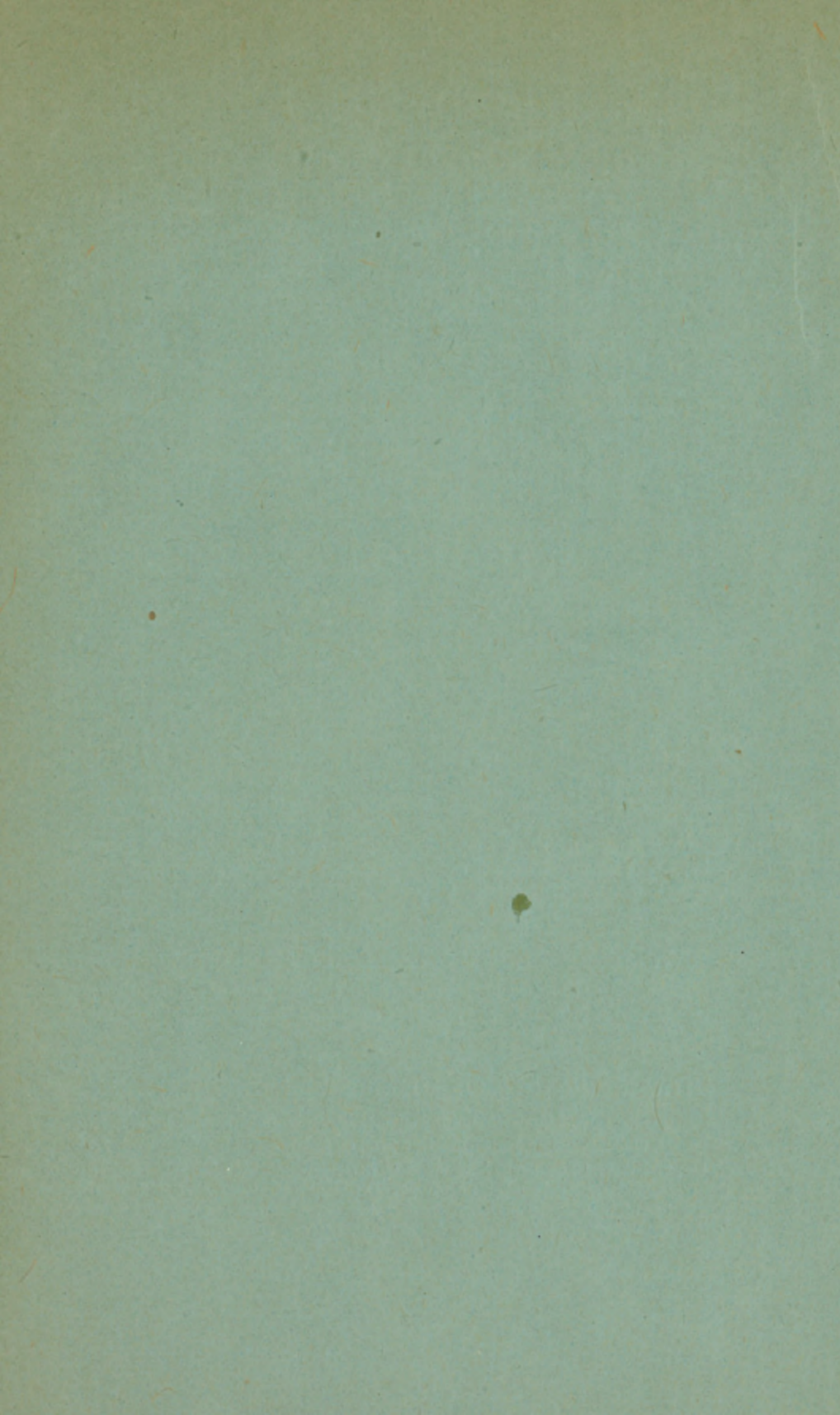
careful study of this condition described by Virchow, and has collected the descriptions of all these skulls in the various museums of Europe. He has shown that some of these skulls present a uniform thickening of all the bones, while others have shown an enlargement limited to the bones of the face, the cranium escaping. He points out the fact that the latter class of skulls probably belonged to patients suffering from the disease acromegalia, in which, as is well known, the bones of the face are very markedly enlarged. It is evident from Baumgarten's study that the term *leontiasis ossea* must be limited to the cases in which the hyperostosis of bone has affected the bones of the cranium only, or these, together with the bones of the face. He considers that the pathological changes present are either a thickening of the surface of the bone alone, or a change both in the surface and the diploe; the latter being in some cases transformed into a spongy tissue; in others, into a hard tissue like ivory. He admits that the literature does not contain any description of this disease observed during life, and he does not think that cases have been observed.

It is my opinion that the case here described is this disease, and that we can affirm that in these cases there is not only a hypertrophy of the bony tissue, but also of the soft parts about it. It is probable that the diffuse thickening of the bones of the head and neck causes some pressure upon the contents of the skull and upon the nerves at their exits between the vertebræ. This might explain the symptoms which are present in our patient—especially the complaints of numbness and the difficulty in her gait.

Various forms of treatment have been pursued in the case under consideration. A thorough trial of thyroid extract failed to affect the condition in any way, and did not cause any variation in temperature from normal. The use of arsenic and strychnine, given in moderate doses, appears to have had some tonic effect, and these, together with the moderate use of corrosive sublimate internally, are the remedies relied upon.

I have ventured to give the condition the name *megalo-cephalie*, because, as already shown, the term *leontiasis* has been applied to three entirely distinct conditions, and therefore leads to confusion.





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