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—BY—

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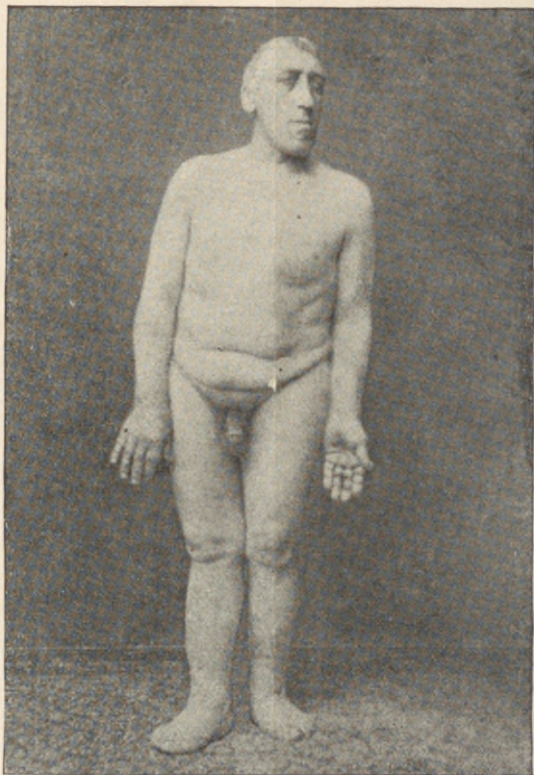
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DR. LONG'S CASE OF ACROMEGALY.

W. H. Lippincott

A Case of Acromegaly.

By CHAS. LONG, A. M., M. D., Wilkes-Barre, Penna.

Symptoms which resemble those of this disease have been grouped together as early as the year 1772, by Sancerotti, and since then various observers have published what were probably cases of this affection under the names, myxoedema, gigantism, exophthalmic goitre, etc. Indeed this group of symptoms was not classed together as a distinct morbid entity until the year 1885, when two cases came under the observation of Marie, then chief assistant to Prof. Charcot, who made a special study of the affection and described its typical symptoms. In naming it, he selected its most prominent symptom, "a striking non-congenital hypertrophy of the extremities (hands, feet and cephalic extremity)" and called it Acromegaly (akro~~a~~ extremity, megas, large). Broca describes it as a disease in which "the bones of the extremities and the extremity of the bones are enlarged."

In Brain, July, '89, Marie gives a biographical index of cases which have come under his notice. This includes eighteen (18) cases of undoubted acromegaly, (one of which is Wadsworth's Boston case); four (4) cases, in which details are wanting, but which he classes as acromegaly, and six (6) cases which do not come under the head of acromegaly at all. Since then one European case has been reported by Drs. Holschewinkoff and von Recklinghausen (Virchow's Archiv, January, 1890), and a case is also now under observation in Dublin, but has, as yet, not been reported. In this country the first case reported was by Dr. Wadsworth (Boston Med. and Surg. Journal, January 1, 1885, as a case of myxoedema, but Marie classes it among his eighteen cases of undoubted acromegaly. Adler, of New York (Medicinische Monatschrift, May, 1889), reported a case in a German woman, 34 years of age. The third case was reported by Dr. O'Connor (American Journal of Homeopathy, 1888). The fourth and fifth cases were reported before the Association of American Physicians at the meeting in May, 1890, by Dr. J. E. Graham, of Toronto, and published in the *Medical News*, October 18th, 1890.



As near as can be determined, therefore, from the researches that I have made, the case which I have the honor of presenting to the society this evening is the sixth reported case of Acromegaly in America.

Case.—Matthias M., German, age 48, laborer (until 10 years ago). His father was a strong, healthy man, who never showed signs of any special enlargements and died at the age of 65, after a two days' illness. Patient can give no information about his mother, who died when he was an infant. He has two brothers and three sisters living, and he thinks they are all healthy; at least, they have never presented any abnormality of growth. The patient's growth was normal, nor did he ever suffer from any sickness during his boyhood or youthful days. At the age of 22 years he entered the German army as a cavalryman, and served three years, until he was 25 years old. At the time he must have been healthy or he would not have been accepted as a soldier. During his service he suffered for five weeks with an abscess on the left hip, but fully recovered. In 1870 he emigrated to America, and was then still a well-proportioned man. Soon after his arrival he was married and lived with his wife for fourteen years. The result of this marriage was three children, two of whom died in infancy, the third is a healthy boy, seventeen years old. About twelve years ago he had a serious abscess, on the right hip this time, from which he also fully recovered. He is sure that he has never had any other sickness, and is free from specific taint. During last summer he suffered for several weeks with headache, but never before or since. He has always been more or less constipated. Symptoms of enlargement were first observed, by the patient's wife, in his hands in 1874, when he was 31 years old, and since that period the hypertrophy has been gradually but constantly increasing. The patient is 5 feet 9½ inches in height and weighs 262½ pounds, an increase in weight of seventeen pounds in twelve years.

Present condition.—The skin has a dirty yellow color, is free from eruption and is very loose, so that it can easily be drawn up in folds. The hands, feet and face are very large, out of proportion to the rest of the body. The face has a distinct elliptical shape so characteristic of patients suffering with Acromegaly.

The cranium is normal in size and shape, and covered with a thick, well preserved crop of gray hair. The forehead recedes from very prominent eye-brows. Neither the eye-lids nor the ears are enlarged. The nose is large and broad. The upper lip is not at all enlarged, but the lower lip is very prominent and everted. The cheek bones are prominent, caused probably by the dilations of the maxillary sinus. The lower jaw is enormously hypertrophied and massive, the chin projecting forward, causing a decided prognathism.

The tongue is very large, thick and wide, and always covered with a grayish-yellow coating. Notwithstanding the enlargement of the tongue, articulation is distinct. The neck is short, thick and bulky, and the head is inclined distinctly forward, due to a slight cervico dorsal kyphosis. The larynx is not all enlarged. The thyroid gland is smaller than normal, but both lobes can be felt. The voice is strong, deep and not at all discordant. Indeed the patient can sing quite well.

The hands are very much hypertrophied, especially in their width and thickness. There is hypertrophy of the soft parts as well as of the bones, and this is especially true of the ulnar side of the hand, where there is an excessive amount of soft tissue. The lines in the palm of the hand are very marked. The fingers are characteristically stumpy and sausage shape, thick and rather flat. The joints are large and flat, showing that hypertrophy is more marked at the ends of the bones. Flexion is complete and the patient is able to make a tolerably good fist. The finger nails are broad and short. The wrist is thick and enlarged in proportion to the hand. The forearm shows some hypertrophy, but the arm is small in proportion to the hand and fore-arm. The muscles of the upper extremity, and indeed, in all parts of the body are flabby and soft. The hand, wrist, foot and ankle of the right side are slightly larger than those of the left side.

The thorax is very large and bulky, especially deep in its antero posterior diameter. There is distinct flattening on the sides. The clavicle and ribs show marked hypertrophy, especially at their extremities. The ribs are inclined very obliquely downward and forward. The scapula is also enlarged. There is a peculiar depression at the upper end of the sternum and no especial prominence of the xiphoid appendage,

which is so marked in the majority of the reported cases. There is also no retrosternal dulness; the thymus gland is therefore absent. Breathing is natural. Enlargement of the heart cannot be detected. The pulse is small, soft and compressible. The abdomen is decidedly prominent. There is no enlargement of the genital organs. The sexual appetite, never very strong, is now almost entirely absent.

The thighs are not abnormally large, except the hypertrophy at the ends of the femur. The knee joints are very prominent.

In the legs both the soft tissue and the long bones show hypertrophy, but that of the muscles is not very marked.

The feet are very large, the increase in size being especially marked in the thickness and breadth. The patient is also flat-footed. The toe nails, like the finger nails, are broad and short. The cutaneous sensibility is good, the knee reflex is normal and the nervous system shows no abnormal symptoms.

Excepting vision the special senses are all normal. The patient is completely blind and has been in this condition for more than ten years. This is, I believe, the first recorded case of acromegaly where blindness is complete. The following is the ophthalmoscopic examination, kindly made for me by Dr. L. H. Taylor :

“Matthias M., aet. 48. Acromegaly, 3, 13, '91. Is now totally blind. Does not distinguish difference between daylight and dark. Cannot distinguish bright lamp held in front of him. Pupils moderately dilated. Eye-balls full but not unusually prominent. Left eye, slight divergent strabismus. He has nystagmus of high grade, so that an ophthalmoscopic examination is difficult.

“3, 15, '91. Atropia dropped in diluted pupils and seemed to steady the eye-ball so that an ophthalmoscopic examination could readily be made. Each eye shows chronic optic atrophy, evidently about the same degree in each. Vessels are small and the nerve greenish. Retina not markedly changed. The patient states that he was examined by Dr. Keyser ten years ago for his eye trouble, and pronounced incurable, having, as he states, trouble with the nerve. It is quite possible that the condition of the eye may have been developed independently of the acromegaly, as he has been blind for ten years. He has used tobacco and also alcohol, but he says not to excess.

Schultz (Neurologisches Centralblatt, July, 1889), reports two cases, in one of which the first symptoms of trouble were disturbances of vision ten years before, which in five years developed into complete temporal hemianopsia. At the time the case was reported the patient was completely blind in one eye and nearly so in the other, so that it was probable that a tumor of the pituitary body was present. In Mr. M.'s case there is nothing to indicate a tumor except total blindness, which of course might be due to tumor of the brain, but in such event other symptoms would probably have been manifest."

The patient has the average intelligence for one in his station of life. His appetite for food and drink is not excessive, and patient says this has always been the case.

Urinary analysis was made by Dr. Lathrop, senior resident physician at the city hospital, Urine Sp. Gr. 1028. No albumen. No sugar. Mucus and slight excess of urates were present. The average daily quantity of urine is 52 1-6 oz., but the daily quantity varies from 29 oz. to 84 oz.

In measuring the patient I have followed the plan adopted by Marie, and I have also made use of the metric system, as well as the inch measure, so that comparison of this patient with others could be made. The measurement of the extremities have all been taken on the left side.

MEASUREMENTS :

Length of hand from lower fold of wrist to end of middle finger	210 mm.—8½ in.
Length of middle finger starting from palmar fold to base	98 mm.—3¾ in.
Length of middle finger on dorsal aspect, starting from the base of its first phalanx	130 mm.—4 7-16 in.
Length of little finger, palmar aspect	78 mm.—3 1-16 in.
Circumference of hand without thumb at metacarpal-phalangeal joints	265 mm. 10 7-16 in.
Width—ditto	120 mm. 4 11-16 in.
Greatest thickness of hand at level of thenar eminence	55 mm. 2 3-16 in.
Circumference of middle-finger	93 mm. 3 11-16 in.
" " thumb	96 mm. 3 12-16 in.
" " little finger	80 mm. 3 3-16 in.
" " wrist immediately below extremity of ulna and radius	220 mm.—7 11-16 in.
Circumference of wrist immediately below styloid proces of ulna	

.....	218 mm.—7 9-16 in.
Circumference of forearm at middle	305 mm.—12 in.
“ “ arm at middle	331 mm.—13 in.
Length from iliac crest to summit of the head of fibula	552 mm.—21 3/4 in.
Length from summit of head of fibula to tip of external maleolus	430 mm.—17 in.
Vertical diameter of patella	70 mm.—2 3/4 in.
Transverse	80 mm.—3 3-16 in.
Circumference of thigh at middle	570 mm.—22 7-16 in.
Greatest circumference of calf	453 mm.—17 9-16 in.
Circumference immediately above tip of internal maleolus	330 mm.—13 in.
Greatest length of foot	295 mm.—11 10-16 in.
Circumference over heel and instep	423 mm.—16 10-16 in.
Greatest circumference of foot	319 mm.—12 1/2 in.
Greatest width of foot	119 mm.—4 11 16 in.
Circumference of big toe	136 mm.—5 5-16 in.
“ “ little toe	75 mm.—3 in.
Length from top of forehead to tip of chin	257 mm.—10 1/8 in.
Length from top of forehead to upper part of nasal bones,	85 mm.—3 5-16 in.
Length from upper part of nasal bones to tip of nose	85 mm.—3 5-16 in.
Greatest width of alae nasi	46 mm.—1 13-16 in.
Distance from tip of nose to point of junction of latter with upper lip	35 mm.—1 6-16 in.
Length from septum of nose to point of chin	103 mm.—4 1-16 in.
Greatest distance between the outer surfaces of the cheek bones	135 mm.—5 5 16 in.
Width of mouth	66 mm.—2 10-16 in.
Vertical measurement of lower lip	16 mm.—10-16 in.
Transverse measurement at middle of tongue	70 mm.—2 3/4 in.
Length of one of borders of tongue pulled out as far as possible to the upper lip	70 mm.—2 3/4 in.
Lower jaw, vertical measurement from free borders of gum to lower part of symphysis	48 mm.—1 14-16 in.
Distance from temporo malar articulation to lower part of symphysis of chin	178 mm.—7 1-16 in.
Distance between the angles of lower jaw	131 mm.—5 3-16 in.
Distance between angles of lower jaw with tape measure, along body of bone passing in front of symphysis	275 mm.—10 13-16 in.
Circumference of thorax over nipple	1230 mm.—48 1/2 in.
Antero posterior diameter of thorax	359 mm.—14 1/8 in.
Circumference of neck between hyoid bone and upper part of thyroid cartilage	440 mm.—17 5-16 in.

(Where there was a projecting body between two points of measurement callipers were employed so as to be accurate.)

There have been so few cases of acromegaly reported that it would be difficult to describe all the typical symptoms, no two cases being exactly alike. The elliptical shaped face and the hypertrophy of the hands and feet, however, have been present in every case.

There is a notable absence of subjective symptoms in Mr. M.'s case. He has suffered slightly with headache, has had two abscesses, and has lost the sexual appetite. He is also completely blind; but Dr. Taylor's report would lead us to infer that the blindness is due to causes distinct from the acromegaly, while he recognizes the possibility of its being caused by hypertrophy of the pituitary body.

There is no constant headache, no polydipsia, polyphagia, or polyuria. The thymus gland is not present, and there are no varicose veins; all of which symptoms have been present in the majority of patients.

The etiology of the disease is unknown. Several of the patients have been syphilitic, but whether this has any effect upon the causation of acromegaly is doubtful. There has never been any proof of heredity.

Acromegaly occurs with about the same frequency in both sexes. It first appears between the ages of 20 years and 30 years; is very slow in its progress, lasting 20 or 30 years or more.

Little is known regarding its morbid anatomy. Marie (Brain, July, '89,) in concluding his description of acromegaly says, "That hypertrophy of the pituitary body with enormous dilatation of the sella turcica, persistence of the thymus, and finally hypertrophy of the cord and ganglia of the sympathetic system not only occur with a remarkable degree of frequency, but may even be looked upon as constant."

That these conditions are not always present is proven in the case reported by Fränzel (*Deutsche Medicinische Wochenschrift*, '88,) and on which an autopsy was performed a year later by Virchow (*Berliner Klinische Wochenschrift*, February 4, '89). Virchow noted absence of the thymus gland, and a normal condition of the thyroid and pituitary glands.

Broca (*Archives Generales de Medicine*, December, '88,) describes the skeleton of one of the original cases of acromegaly by Marie. "The principal change found was an hypertrophied and increased porosity of the bones of the extremities

and enlargement of the channels containing blood vessels. There were also exostoses at the articular extremities of the bones. The inferior maxillary bone was greatly hypertrophied, and all the sinuses in the bones of the skull were dilated."

Diagnosis.—The case presented cannot well be confounded with any of the diseases which resemble Acromegaly.

In myxoedema the soft parts alone are affected. The skin is thickened and scaly, and both it and the subcutaneous cellular tissue present a waxy like condition. The face is round and the mental condition is bad. The skeleton, on the other hand, is unaffected. The symptoms are all absent in the patient presented.

The Leontiasis ossea of Virchow gives rise to bony tumors on the face and cranium, causing a hideous appearance and great deformity.

Osteitis deformans of Paget generally manifests itself after forty years, whereas Acromegaly appears from ten to twenty years earlier. The deformity does not occur symmetrically, as it does in this patient, and hypertrophy affects the long bones in preference to the bones of the extremity and the extremity of the bones. The long bones also present marked deformities. The bones of the cranium also undergo hyperostosis instead of the bones of the face, giving the latter a triangular and not an elliptical shape.

In gigantism all parts are proportionately enlarged, there being no special hypertrophy of the lower jaw, hands and feet.

Marie will not admit that Friedrich's disease, in which hypertrophies occur somewhat similarly to those of Acromegaly, has any connection with this disease, and also will not allow it to be confused with unilateral hypertrophies, which sometimes occur in different parts of the body.

Treatment has had no effect upon the progress of this disease. In the present case there has been no attempt at treatment.

My thanks are due to Dr. G. W. Guthrie for allowing me to observe the patient in the City Hospital; Dr. L. H. Taylor for the ophthalmoscopic examination, and Dr. Lathrop, (Resident of the Hospital), for the urinary analysis.

