

CHURCH (A.) & HESSERT (W.)

# ACROMEGALY

*WITH THE CLINICAL REPORT OF A CASE*

By

ARCHIBALD CHURCH, M.D.

PROFESSOR OF NEUROLOGY IN THE CHICAGO POLYCLINIC; LECTURER ON INSANITY IN THE CHICAGO MEDICAL COLLEGE; NEUROLOGIST TO THE COOK COUNTY HOSPITAL, CHICAGO, ILL.

AND

WILLIAM HESSERT, M.D.

RESIDENT STAFF OF COOK COUNTY HOSPITAL, CHICAGO, ILL.

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## ACROMEGALY, WITH THE CLINICAL REPORT OF A CASE.

SOME years ago, Dr. Paul Marie, at that time Chief of Clinic under Professor Charcot, described a peculiar condition presented by two patients in the service, consisting in a notable symmetrical enlargement of the extremities and the face. This description,<sup>1</sup> which is classical and to which nothing material has since been added, although at present something like ninety cases from various sources have been recorded, led to many subsequent observations and to a number of theories as to the etiology of the disease. An important communication by Souza-Leite<sup>2</sup> gave abstracts of forty-nine cases, and was translated into English and published by the Sydenham Society. Recently, in the *Journal of Nervous and Mental Diseases*, December, 1892, the literature of the subject has been brought up to date by Dr. Joseph Collins, of New York, who gives a digest of all cases published subsequent to Souza-Leite's report and previous to June, 1892. Since that time perhaps a dozen have been recorded, and though the condition at first was considered absolutely rare, as in other similar instances, the diffusion of knowledge concerning it has led to the recognition of the disease in all directions. Dr. Collins also gives the bibliography of the subject in the same journal for February of this year, to which we are much indebted.

The disease presents, except in rare instances, no trace of heredity, but for the most part the subjects of this mal-

<sup>1</sup> *Revue de Médecine*, 1886.

<sup>2</sup> *De l'Acromégalie*, Paris, 1890.

ady, at least in this country, have been in such a position that it is impossible to trace their lineage with any satisfaction. It appears usually at puberty or at the age of complete sexual development, between the twelfth and thirtieth years. In some instances it has seemed to be congenital, and in others it has appeared late in life. Among the cases so far on record there is a slight preponderance of males. As inciting causes, such a variety have been alleged or suspected that weight attaches to none.

Usually the hands commence to enlarge, and shortly the feet present a similar growth, followed in turn by changes of a hypertrophic nature involving the facial bones and soft parts, particularly the region of the frontal sinuses and the under jaw. Occasionally the bones of the thorax present similar features, and not infrequently the spine becomes scoliotic, presenting in marked cases a notable antero-posterior curvature in the upper dorsal region, giving rise to a well-marked hump. The involved tissues have a peculiar resilient, non-œdematous feel, do not pit on pressure, and palpably manifest the enlargement of the underlying bony structures as well as of the soft parts. The overhanging brow, the prominent malar eminences, the thickened nose and lips, the protruded and greatly enlarged chin, with the heavy expression and the exaggeration of the more prominent facial folds, gives a characteristic appearance to these patients which once seen can scarcely be subsequently misconstrued, and reminds one somewhat of the leonine expression that goes with leprosy, though more mask-like and expressionless. The scalp and ears frequently show similar changes, and many of these patients require to increase the size of their hats from time to time. Examination of the mucous membrane of the naso-pharynx; the fauces, and the mouth discloses a somewhat similar condition, and the low, guttural voice, as well as the actually increased size of the larynx indicates its hypertrophic involvement. Owing to the enlargement of the bony arches of the jaws, intervals appear between the teeth, which latter of course do

not correspondingly increase in size. The tongue becomes voluminous and frequently shows on its dorsum, by the hypertrophy of the mucous structure, a rugous, corrugated appearance. In the case we have to report the tongue, when protruded and spread out, nearly covers the enormously hypertrophied chin, and in some instances reported it has been possible for the patient to lap the end of the nose with the tip of the tongue. The bones of the shoulder girdle, notably the clavicles, sometimes participate in the hypertrophic process to an extent that causes a broadening of the shoulders. The arms as a rule escape, though there may be slight enlargement at the elbows. But beginning with the middle of the forearm and extending to the tips of the fingers, the enlargement is pronounced, giving rise to the descriptions of these extremities as being battledore-shaped or spade-like. As the hypertrophy increases the strength diminishes, yet flexibility and precision of movements and co ordination are not affected. Sexual power is usually much diminished or entirely lost at an early period. On the palmar surface of the hand the integument is notably thickened and the ordinary folds become enormously exaggerated. The fingers, thickened in all directions, are well described as sausage-shaped; the nails correspondingly increase in size, but rather in width than in length, and frequently are coarse, thick, striated, and roughened. Indeed, all the dermal structures in the hypertrophied territory manifest this trophic disturbance by excessive growth, the cuticle becoming rough and thickened, the hairs harsh and bristly. In the feet and lower legs similar changes are produced, giving rise to greater length, width, and thickness of the feet, which present usually on the inner side, but sometimes on the outer margin, a peculiar thickened welt encircling the heel and running down the margin of the foot to the corresponding phalangeal joint. The toes show similar changes to those of the fingers.

In some cases, as first pointed out by Klebs,<sup>1</sup> there is a

<sup>1</sup> Die krankhaften Störungen, etc., 1889.

persistence of the thymus gland, which has been found once or twice post mortem, and suspected in some other cases by the post sternal dulness. Rarely the thyroid has been found enlarged, but as a rule, both during life and post mortem, it is notably diminished, and in some cases practically wanting. The spleen has been found hypertrophied, and all glandular structures, especially the ductless glands, have been found variously involved in the direction either of hypertrophy or of atrophy. This is particularly true of the glandular portion of the pituitary body, which, in about fifty cases, has been found either on post-mortem examination or from symptoms noted during life, to be greatly enlarged. This enlargement bringing pressure to bear upon the optic tracts and chiasm, or upon the optic nerves, results in more or less loss of the visual field, which may be confined to the temporal portions owing to the involvement of the nasal half of the retina, and this is the most common limitation; or it may show itself by a concentric or hemiopic limitation of the field. Hearing is not rarely involved. In some cases the sense of smell has been lost, and a peculiar mental hebetude approaching idiocy has been frequently noted. A very fair proportion of the cases present symptoms, on the part of the urine particularly, in the direction of glycosuria and polyuria with attending polydipsia.

Some cases run a comparatively rapid course, terminating in a few years, while others present a very protracted history, the disease not apparently shortening life.

Although numerous theories have been propounded explanatory of this diseased condition, none of them is as yet accepted without qualifications. It was the idea of Marie,<sup>1</sup> though not categorically stated, that the involvement of the pituitary body was the essential feature. Klebs<sup>2</sup> maintained that the persistence of the thymus gland was of the utmost significance. Virchow<sup>3</sup> believes that acromegaly is merely the terminal stage of a condi-

<sup>1</sup> Loc. cit.

<sup>2</sup> Loc. cit.

<sup>3</sup> Berliner klin. Wochenschrift, 1889.

tion the early periods of which are not yet recognized. By others it is conceived that the disease is neurotic, depending either upon the presence in the blood of a certain substance necessary for proper trophic action, which is supposed to be the secretion of the pituitary body; or, on the other hand, to the presence in the blood of poisonous products which result in dystrophy, and which normally are eliminated by the pituitary. This theory, of course, is parallel to the one erected by Horsley, expounding the relation of the thyroid body to myxœdema. The interrelation between the thymus, the thyroid, and the pituitary bodies is shown clinically in such conditions as myxœdema and cretinism, and experimentally by some observers who have found in animals, that where the thyroid has been extirpated the prehypophysis cerebri has shown hypertrophic enlargement. It must be added that certain cases have been reported in which the pituitary body has been enlarged to an enormous extent without attending symptoms of acromegaly, and still other cases in which it has been absolutely destroyed by disease, and acromegaly has not been induced. It is difficult, however, to see why the dystrophy originating from a cause such as has been suggested, should be practically confined to the extremities, as such dystrophy must be dependent for its mechanism upon a change in the vaso-motor control through which nutrition is regulated. One or two instances are recorded in which what is denominated hypertrophy of the sympathetic nervous system has been alleged, and there are those who are inclined to attribute to this change in the sympathetic system the peculiar conditions found in this disease.

Inasmuch, however, as the sympathetic system cannot be dissociated from the cerebro-spinal axis, it is scarcely wise to attribute to its involvement the rôle in question.

In some few cases the blood has been investigated without the detection of any peculiarities of a suggestive nature. Our own impressions are that the disease is a near relative of cretinism and myxœdema, to which it

shows in certain instances a very close resemblance. With this idea in mind it is our intention, at the first opportunity, in a comparatively early period of the disease, to attempt treatment by the use of thyroid juices administered either hypodermically or otherwise; a method of treating myxœdema which has received the very strongest support from the results obtained, especially by British observers. All other means of treatment for this condition hitherto attempted have apparently been futile.

A. K.—, aged forty one, single, cabinet maker.

*Family History.*—The patient, being of illegitimate parentage, can supply but a meagre account of his ancestry. Of his father he knows nothing. His mother, with whom he lived until he was seven or eight years of age, he remembers as a short, stout, heavy-set woman, with no peculiarities or deformities.

*Personal History.*—Is a native of Silesian Prussia, where cretinism and goitre are not uncommon. His previous health record seems free from any disease of consequence, as he has always, until lately, enjoyed the best of health and spirits. He had a mild attack of malaria some years ago. While still a young lad, and on the occasion of his purchase of a pair of kid gloves, his attention was directed to the size of hands, which were disproportionately large, calling for a man's size glove. He does not remember whether his feet were large at that time, as his shoes were made to measure, as is customary in Germany. Nor did anyone remark any peculiarity about his face and head. He never had occasion to consult a physician for illness, and ten years ago, at the age of thirty, he came to America. Here he found that ready-made shoes were difficult to obtain, on account of the width of his feet. It also became apparent to him that he was an object of attention and ridicule on the street, and remarks were passed about his large head and hideous aspect. It has not been necessary for him to obtain a larger size hat, having worn a  $7\frac{3}{4}$  for a long time. Shortly after arrival in New York he became infected with syphilis, and in due



course of time had mucous patches and warts about the genitals and anus. He denies ever having had any skin eruptions, alopecia, sore throat, or iritis. During the last few months a crop of tertiary lesions has developed on the skin. Sexual desire and ability are apparently retained. The appetite is voracious. For the last three years he has been obliged to satisfy a steadily increasing thirst with large quantities of water or beer. Urination has been growing more frequent, and he rises a few times at night to void large amounts of urine. Until about five years ago he had been of a cheerful disposition and mentally active for one of his class, but since that time he is subject to fits of melancholy, at times accompanied by suicidal thoughts.

His memory and mental faculties are becoming weakened so that he cannot pursue a connected train of thought, and says he is at times quite "befuddled." He sleeps much and dozes off at any time of day. He has felt for some time that he is losing ground both physically and mentally, and his physical condition and general hideous aspect are such that he can with difficulty find employment. He has slight headaches oc-

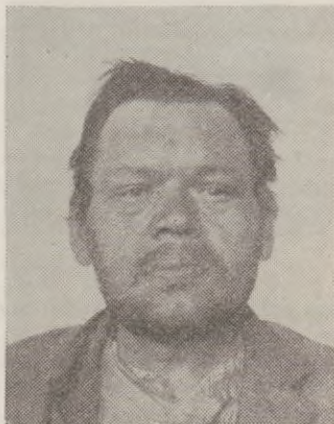


FIG. 1.

asionally, and is subject to nasal catarrh. He applied at the hospital for treatment for the syphilitic trouble.

*Present Condition.*—The patient is small of stature, height, 5 feet  $4\frac{1}{4}$  inches, weight, 179 pounds. His gait and movements are steady and co-ordinate. Attention being first directed to the head (Figs. 1 and 2),

one is struck by the coarse features. The hair is fairly heavy, beard sparse, short, and stubby. The forehead is broad and receding, the superciliary ridges and frontal sinuses are enormously developed, the malar bones are

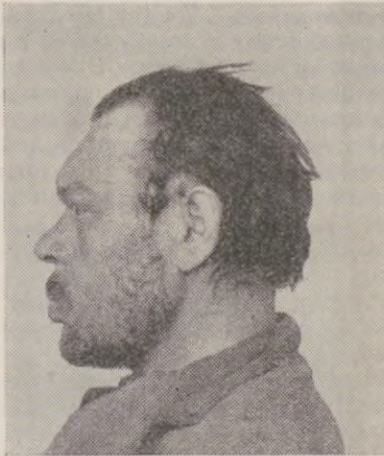


FIG. 2.

prominent, the eyelids are somewhat thick and heavy, the pupils are equal in size and react the light. The nose is large and broad, overhung by the massive brows, the alæ nasi are thick, and the nostrils large. The ears are very prominent, but do not project excessively from the sides of the head. The lower jaw is very heavy and massive, especially the rami, the chin projects but slightly,

and prognathism is not pronounced. The lips are thick and everted, especially the lower. The teeth are somewhat decayed, and in the lower jaw marked interval exists between them, but the lower teeth do not project beyond the upper teeth. The tongue when protruded almost touches the point of the chin, and is broad and thick. The face does not present an elongated oval, but is rather broad laterally. The throat and naso-pharynx are apparently negative. The left nostril is somewhat occluded by the swollen turbinated body. The voice is harsh, low, and guttural, the neck is short and thick. The thyroid cartilage is normal in size, but the thyroid gland can be distinguished with difficulty, being apparently diminished in

size. There are a few warty growths on the sides of the neck. The thorax is of normal dimensions, there being no increase in the shoulder girdle, nor is there any bony hypertrophy. The heart and lungs are normal, and the post-sternal dulness of Erb is not demonstrable. The abdomen is very prominent, flabby, and pendulous. Examination of the abdominal viscera gives negative results. There is no cervico-dorsal kyphosis nor any lateral deviation. The muscles are under-developed, soft, and flabby. The strength in the arms is much diminished, as tested



FIG. 3.

by the dynamometer, being twenty for the right and thirty-eight for the left hand.

Interest next centres on the hands, which are in striking contrast to the forearms, whose lack of muscular curves is thereby rendered more notable. The wrists are

but slightly thickened, but the hands are uniformly and symmetrically enlarged (Figs. 3 and 4). The integument on the palmar surface is very thick, inelastic, calloused, and the normal furrows and markings are deeply exag-

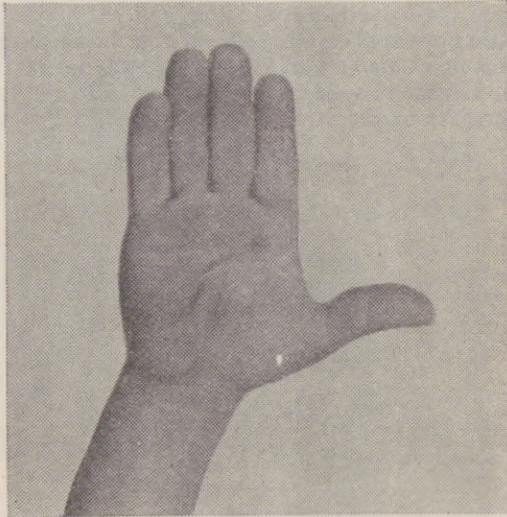


FIG. 4.

gerated. The fingers and thumbs are relatively short, stumpy, broad, and flat—"sausage shaped." The nails are broader than long, flat, not beaked or fissured, and appear to be imbedded in a superabundance of tissue. The hand measures  $4\frac{1}{2}$  inches in width and 8 inches in length.

The feet (Fig. 5) are enormous, and measure  $4\frac{1}{4}$  inches in width and  $10\frac{3}{4}$  inches in length. The toes, like the fingers, are broad, short, and thick, and preserve their relative proportions, the nails being flat and overhanging by tissues. The calcaneum projects markedly, and

a large bunion is conspicuous. A prominent feature about the member is a welt or pad of hard superabundant tissue placed on the outer and posterior aspect of the foot, contributing much to its increased size, and at the hallux recalling the heel of the negro. The ankles and legs are not appreciably enlarged. There is no implication of the joints anywhere, and all movements are free and accurate.

*Skin.*—There is a tertiary syphilitic eruption distributed asymmetrically over the trunk and extremities,

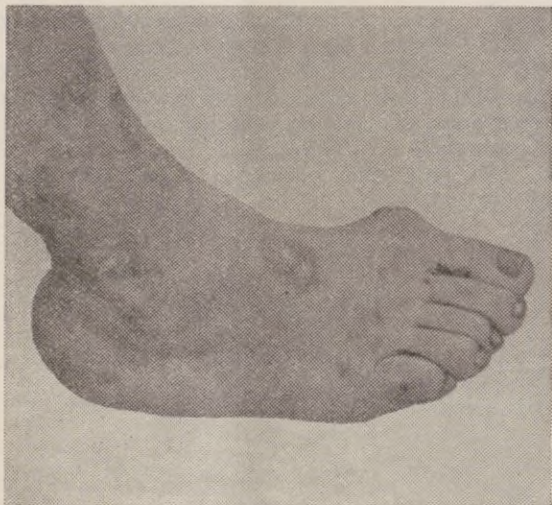


FIG. 5.

especially over the legs and feet, consisting of atrophic pigmented scars, and papulo-squamous and crustous lesions. The post-cervical and cubital glands are somewhat enlarged. There is no œdema at any point.

*Genito-urinary System.*—The total daily amount of

urine is high. No albumin; sugar, 6.67 per cent. No difficulty in urination. The penis is of average dimensions, the scrotum is loose and long, and the testes are rather large.

*Special Senses.*—There are no sensory disturbances either objective or subjective; no cephalalgia nor neuralgic disturbances. The patellar reflex is sluggish. There is no vertigo, loss of consciousness, or grave emotional disturbances. Hearing and smell are fairly acute; but there is myopia of high degree. There is some diminution of the field of vision on the temporal side for the right eye. The red color field is diminished in both eyes. The retina and nerve seem normal. Taste is normal.

*Blood.*—Examination of the blood shows the amount of hæmoglobin to be ninety-five per cent. of the normal, and an average of ninety-six countings with the hæmocytometer shows seven million red corpuscles to the cubic mm. The proportion of white to red corpuscles is about 1 to 400.



