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ON ACROMEGALY AND GIGANTISM, WITH UNILATERAL FACIAL HYPERTROPHY;—CASES WITH AUTOPSY.¹

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IN the present paper I have placed together the report of a case of acromegaly occurring in a professional "giant," and the history of another giant with some symptoms of acromegaly, and a very striking form of progressive unilateral facial hypertrophy.

CASE I.—GIGANTISM AND ACROMEGALY.

The first patient, Santos Mamai, was a Bolivian Indian, a man thirty years of age, who was exhibited in this city under the name of the Peruvian giant. He was advertised to be seven feet eight inches high and to weigh 330 pounds; in fact his height was six feet seven inches, and his weight about 300 pounds. Absolutely nothing could be learned by me regarding the history of the patient, except that he came to this country last fall with a troupe of other Bolivian Indians who all claimed to be lineal descendants of the Incas. Their object was to exhibit themselves; but they did not succeed, and becoming

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"stranded," the troupe was brought to this city, where Santos was taken ill. He was said to have been a person of very quiet and even melancholy temperament, reticent, rather feeble in mind, and perhaps very homesick. Although of gigantic proportions, he was not very strong, nor was his muscular development great. Without any known cause, he was suddenly taken very ill and brought into my wards at Bellevue Hospital.

On admission he was unable to stand or sit, owing to excessive weakness. His mind was very dull and irresponsible. His pulse was feeble and rather rapid, 108. Respiration loud, 24 per minute, harsh and labored. There were some physical signs of a bronchitis. The temperature was normal. He apparently was suffering from no pain, and presented no evidence of any paralysis or any manifestly acute disease; he simply seemed to be in a state of collapse. In spite of stimulants this collapse increased, and he, in the course of four or five hours, passed into a state of coma and remained so until death, about twelve hours after admission.

The physiognomy of the man, when first seen by me, suggested at once the possibility of his being a case of acromegaly, and measurements of the body were taken accordingly. These measurements, with a description of the person and the post-mortem findings, convinced me that the diagnosis is correct. His height was, as stated, six feet seven inches, and the weight 300 pounds. The most striking appearance, however, was the very large under jaw and the enormous thorax. The measurement from the angle of the lower jaw to its symphysis was $14\frac{1}{2}$ cm. or $5\frac{3}{4}$ inches; the average measurement of an adult man being not over 10 cm. or $3\frac{3}{4}$ inches. The length of the face from the root of the hair to the chin was 34 cm. or $13\frac{1}{2}$ inches; from the crown of lower incisors to the point of the chin 7 cm. or $2\frac{3}{8}$ inches. These measurements indicate that the man had a disproportionately developed face, the enlargement affecting particularly the lower jaw. The malar bones were very prominent, as is usual with the Indian. The circumference of the head was 56 cm., not being very greatly in excess of the average. The circumference of the thorax was 50 inches, as against the average for the adult man of 34. This enormous development of the thorax was, together with the enormous enlargement of the face, the striking feature of the case. The man had large hands

and large feet, somewhat in excess proportionately of what they should be. The nails were normal. The ears and tongue were of normal size. There was some kyphosis. On making the autopsy, the scalp was found to be excessively thick and lying in folds as though the skin had grown and was intended for a much larger skull. The hair was excessively coarse and thick. The special measurements of the different parts of the body are given below.

AUTOPSY.

The autopsy was made by my house physician, Dr. C. J. Strong, and I am indebted to him and to Dr. McAlpin for the notes as to the condition of the body. Unfortunately the autopsy was not made with reference to the possibility of the case being one of acromegaly, and some details were therefore incomplete.

Muscular development very poor.

Heart.—Weight, 23 oz. Thickened patch on pericardium. Mitral valve admits two fingers; aortic valves normal. Muscular fibres coarse, with considerable connective tissue between them. Tricuspid valves normal. Hypertrophy more marked in right heart.

Lungs.—Left, weight 20 oz. Pleural congestion, slight interlobar pleurisy, some emphysema. Moderate amount of passive congestion. Right, weight 27 oz. Pleura adherent to diaphragm. Considerable thickening of the interstitial tissue. Both very light in color. Almost total absence of anthrakosis.

Spleen.—Weight, 7 oz. Passive congestion. Substance fine dark chocolate color.

Kidneys.—Weight, left 14 oz., right 13 oz. Left cortex swollen, somewhat cloudy. Markings coarse and irregular. Capsule very thin and strips off easily. Malpighian bodies show plainly on account of congestion. Right same as left.

Liver.—Weight 8 lbs. 3 oz. Large and fatty. Margin sharp and well defined; substance pale yellow. Lobules hard to make out; soft; opaque color; cloudy swelling of epithelium of lobules.

The thyroid gland weighed 4 oz. and had a perfectly normal appearance.

The brain was dry, pale and firm; its weight was 53 oz. There were no evidences of exudation or inflammation. The blood-vessels were normal. On removing the brain it was noticed that the *pituitary gland* was

very much enlarged, and in dissecting it out of the sella turcica a part of it which seemed to be firmly attached to the bone was torn, a slight amount of blood and serous fluid exuding. It was somewhat spherical in shape, and showed at its inferior surface the line defining anterior and posterior parts. The transverse and antero-posterior diameters are each about 3 cm. The weight of the gland was 4.5 grammes. It was attached to a pedicle that was 1.5 cm. long. The gland was of rather soft consistency and apparently somewhat cystic. The tear in the

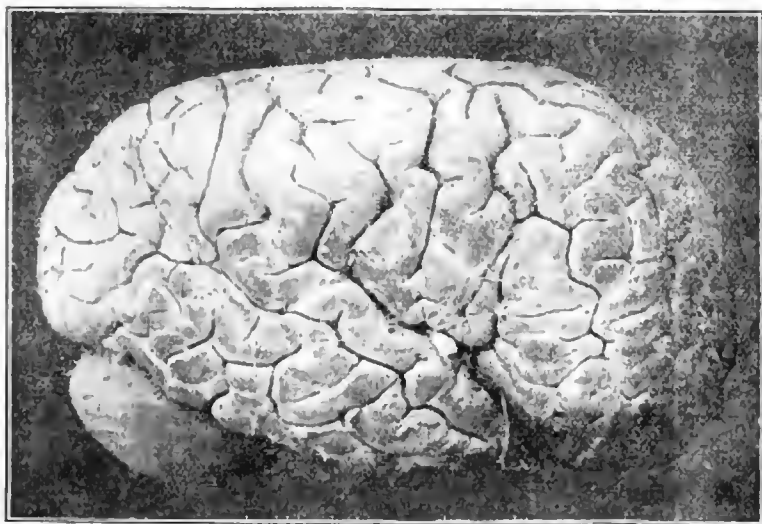


FIGURE I. Brain of Peruvian Giant, one-half actual size.
Left Hemisphere.

anterior portion had reduced somewhat its bulk by reason of the loss of hemorrhagic contents. All other parts of the brain appeared perfectly normal.

A study of the convolucional development was made, and some measurements and notes taken. These have only an anatomical and anthropological interest; they will be given in a later paper in connection with the photographs. The special measurements of the body were made by me with the help of Dr. Stivers, who has furnished me with a copy of the notes.

BODY MEASUREMENTS OF GIANT.

Height, 6 feet 10 inches.

Supposed weight, 300 pounds.

HEAD.—Greatest circumference at level of glabella,	55	cm.	22	in.
Distance between external orbital processes,	21.5	cm.		
Width of face at level of zygomas,	17	cm.	$6\frac{3}{4}$	in.
Root of hair to point of chin,	24	cm.	$9\frac{1}{2}$	in.
Angle of inferior maxilla to its symphysis,	14.5	cm.	$5\frac{3}{4}$	in.
Width of mouth,	7.7	cm.		
Thickness of lower lip,	1.75	cm.		
From crown of lower incisors to point of chin,	7	cm.	$2\frac{3}{4}$	in.
Length of nose,	7	cm.		

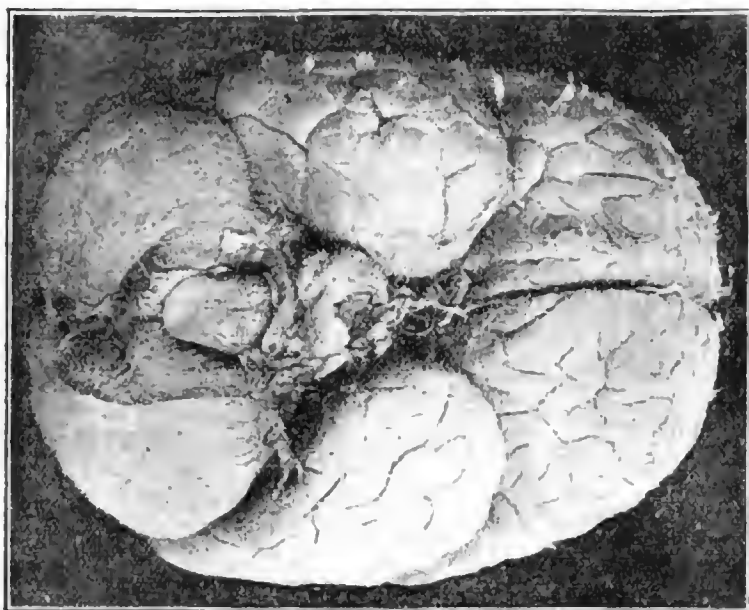


FIGURE 2. Same, showing enlarged pituitary body.

BODY.—Circumference of chest,	126	cm.	50	in.
Circumference of waist,	104	cm.		
UPPER EXTREMITIES.—From tip of acromion process to olecranon (upper arm),	41	cm.		
From olecranon to styloid pro- cess of radius (lower arm),	34	cm.		
Length of hand from tip of middle finger to wrist,	22	cm.	$8\frac{5}{8}$	in.
Length of first finger,	13	cm.		

Length of second finger,	15	cm.
Length of third finger,	14	cm.
Length of fourth finger,	12	cm.
Circumference of fingers (first joint) ranges from 7.5 to	8.5	cm.
Circumference of wrist,	21	cm.
The fingers were not spade-like.		

LOWER EXTREMITIES.—From crest of ilium to upper edge of patella,			51.5	cm.
From upper edge of patella to sole of foot,			63	cm.
Total length of foot,			29	cm. 11½ in.

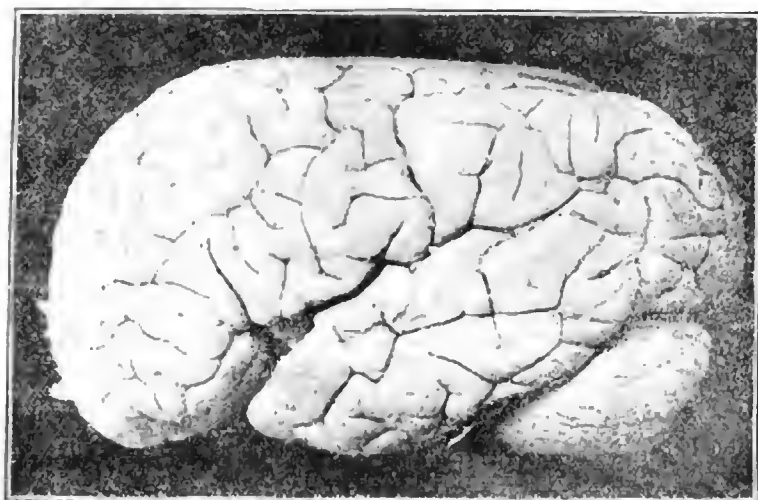


FIGURE 3. Same, right hemisphere.

Length of toes range from 5.5 to	7.5	cm.
Circumference of ankle,	25	cm.
Circumference of foot at instep,	33	cm. 13 in.
Circumference of foot at ball of foot,	30.5	cm.

There is a considerable cushion or pad of flesh on the outer side of each foot.

Distance between anterior superior spinous processes of the ilia,	37	cm.
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The penis was extremely small, measuring 3 in. in length.

REMARKS.

The diagnosis of this case is based upon the enormous development of the face, particularly of the lower part; the enormous development of the thorax; the hypertrophy of the skin of the scalp; the somewhat disproportionately enlarged extremities; the cushion of flesh on the outer side of the soles of the feet; and consequent thickness of the feet; the history of the patient, which shows a person of enormous size and very deficient

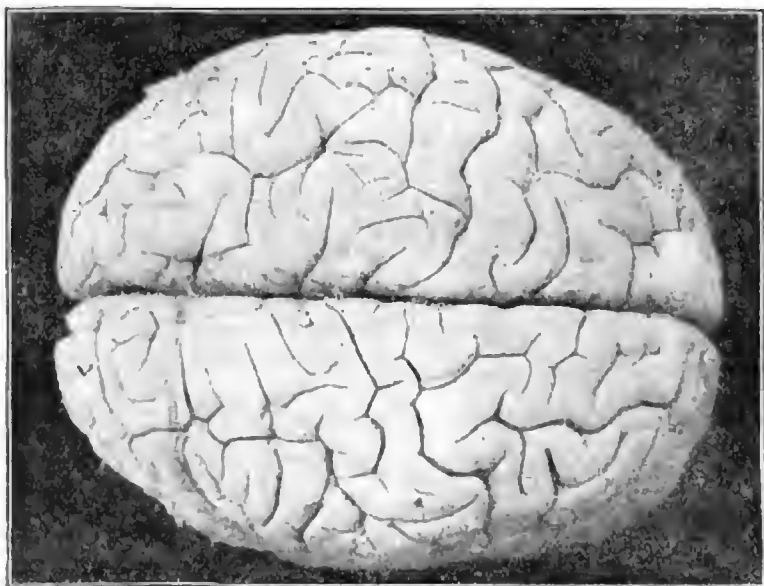


FIGURE 4. Same.

muscular strength, with some mental weakness; and finally the existence of an enlargement of the pituitary gland. Whether the pituitary gland is the cause of the disease or not I am unable to say, and shall not venture upon an opinion. The evidence, however, which places an enlargement of this gland into relationship with the condition known as acromegaly is certainly increasing. There have been, so far as I can find, only eleven autopsies made among the eighty odd cases reported.

In nine of these an enlargement of the pituitary body was found. This enlargement was very much like that in my case, in four cases, viz.:

In Holsti's case the measurement was 2.5 x 3.0 cm.; Hensot, 3.0 x 4.2; Brigidi, 2.8 x 3.8; Heron, 3.2 x 3.8. In Taruffi's skeleton, and in one of Marie's cases, the enlargement was greater. In the two cases in which there was no enlargement, the symptoms ran an acute and peculiar course. In one of them there was atrophy and paralysis of the arms, with symptoms of bulbar palsy. Post-mortem: several softened cavities were found in the brain. This may have been a case of glomatosis (*Brit. Med. Journal*, Vol. 1890, p. 662). In the second case there was a sarcoma found in the lungs (*Illustrated Med. News*, 1889, Vol. II, p. 195). The fact that the gland sometimes does become enlarged or diseased without producing acromegaly does not necessarily militate against the theory of its relationship to acromegaly, for it may be only a peculiar perversion of the function of the gland which can produce symptoms of the disorder. Furthermore, it is extremely probable that the pituitary gland, if it has any functions, has them in connection with other glands, and that when its function is abolished other glands can in some cases take its place. Thus we find that adults can live very well for a considerable period without any spleen. In my case, the actual note with regard to the condition of the thymus was not made; but the autopsy was a very careful one, and I am sure if that gland had been present it would have been noticed. The thyroid gland appears to have been normal in size and in anatomical structure; it does not seem to me that there is any evidence whatever that this gland has a relation to acromegaly.

The discovery and description of acromegaly, and pathological studies that have been made upon it, have done at least this good: they have brought into evidence the pituitary body and have led to much careful investigation of its functions. This is a body which has before been considered vestigial in character, but it is now

coming to be regarded as an organ of some importance. In a very careful study of it by Dr. Boyce (*Journal of Bacteriology and Pathology*) a number of interesting facts with regard to it have been collected. Dr. Boyce examined and weighed the pituitary gland in over 100 cases. The patients were insane and had died from intercurrent diseases. He found the average weight to be from 0.3 to 0.6 gm., the average being 0.5 gm. He found that this weight bore no relation to age and none particularly to sex or to general nutrition or to the size of the brain. He reports one case in which the gland was absent in a person who died from phthisis. He gives a summary of the anatomical studies of the gland and also of the pathological changes which affect it. It is known that the gland is composed of an anterior portion which is related to the alimentary tract and a posterior portion relating to the central canal of the spinal cord. The nervous or posterior portion is decidedly atrophied, and is unquestionably vestigial or rudimentary; but the anterior portion or glandular part may have some special function, and it is this part which seems to be specially subject to disease. It is this part also which was especially diseased in my own case. The gland seems to be somewhat enlarged after extirpation of the thyroid, at least in animals. It is somewhat enlarged in myxœdema and in cretinism; but the enlargement in all these cases seems to be slight and of little consequence. The only disease in which the gland is markedly and strikingly enlarged is acromegaly.

Another point of interest in connection with my case is the question of acromegaly in its relation to giant development. There have been several other cases of acromegaly in which the general stature and size of the person were gigantic. Three of the cases reported by American observers were of persons who were over six feet high.

There is one case like mine reported by Taruffi. The man died in 1808. The skeleton measures 1.8 (six feet in height), and had all the marks of acromegaly.

Furthermore, the sella turcica was so enlarged that it must be assumed that the pituitary body was hypertrophied.

Virchow also reported a case of acromegaly in a man of enormous size and muscular strength. His height was 1.838 (6.1½).

In several other instances the patients have been very large men as shown here:

Cohen's case,	6 feet 2½ inches.	Weight, 238 pounds
Barclay and Sommers,	6 " 2 "	" 280 "
Packard,	6 " 1½ "	" 210 "
Sommers,	5 " 11 "	" 225 "
Long,	5 " 9½ "	" 262 "
Osborne,	5 " 9 "	" 225 "
Alfieri,	6 " 4 "	

My own case was one in which the individual was of usual stature, and it seems to me that it is not unlikely that many of the cases of giant growth on exhibition as such may be cases of peculiar and aborted types of acromegaly. I have made some attempts to study dime museum giants since my case came under observation, but the giant business has been at a low ebb in New York of late, and I have had only one opportunity for observation.

The following case seems to be very apposite in this connection, because together with a gigantic general growth, there is a special hypertrophy of part of one extremity. It might be called a case of *somatomegaly*, a name suggested to me by Dr. Frank P. Foster, with a hemiacromegaly of the head. In plainer language, the individual is a professional giant with an enormous special development of one-half the face.

CASE II.—Lewis Wilkin, aged 19; single; occupation freak; was born in Minnesota. His father was a native of New York, his mother of Canada, of English stock. His parents were healthy people, of average size. He has six brothers and sisters, all of ordinary height. He was the second child. He was always large for his age, though not remarkably so. He grew steadily, however,

until by the age of 17, he was over seven feet. He is now nearly 20 years old, and has grown one and a half inches in the last year. His present height is seven feet four inches. His weight 325 pounds. His general proportions are for the most part good, but his feet and hands are particularly enormous, and the left side of the face shows a remarkable osseous hypertrophy, involving the frontal bone, upper and lower jaws. The hyper-



FIGURE 5. Gigantism with facial hemihypertrophy.

trophy corresponds closely with the distribution of the left trigeminal nerve. It gives his face a curious twisted and symmetrical look, which is shown imperfectly in the photograph. The first impression is that he has a right hemiatrophy of the face. Closer inspection reveals, however, an enormous thickening of the left upper alveolar processes. The bone bulges out above the teeth as though he had a gum boil. The palatal arch is

also greatly enlarged on the left side. The lower jaw is less affected, but is larger and longer on the left side. The teeth are white and even, and are not enlarged. The orbits are alike, but the left brow, and indeed the whole frontal bone, bulges out so as to give a curious deformity to the skull.

I could not get an exact outline, but it is shown approximately here. The thickening reaches back as far

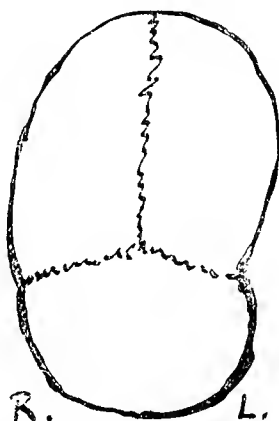


FIGURE 6. Gigantism with hemihypertrophy, showing outline of skull.

as the coronal suture and stops there. The head in brief is large, measuring 65.5 cm. in its greatest circumference. The osseous hypertrophy makes the naso-bregmatic arc very large, viz., 18.5, while the binauricular arc, measured through the bregma, is relatively smaller, as it does not go over the hypertrophied area. The measurements are:

Greatest circumference of head,	65.5 cm.	25 $\frac{3}{4}$ in.
Naso-occipital arc,	43. "	17 "
" bregmatic "	18. "	7 $\frac{1}{2}$ "
Binauricular "	37. "	14 $\frac{3}{4}$ "

From angle of jaw to symphysis of chin, right side, 13. ($5\frac{1}{8}$), left side 18. ($7\frac{1}{8}$), a difference of two inches.

The circumference of the chest, at the mammary line, was 47 $\frac{1}{2}$ inches, and the expansion three inches. This shows that he has a thorax of not excessive size proportionately. It is a good deal smaller than that of the Indian giant, whose height was 6 feet 7 inches, and whose chest measured 50 inches.

The hands were enormous, measuring 26 cm. or $10\frac{1}{4}$ inches from the tip of the middle finger to the process of the ulnar, the circumference of the open hand around the middle of the palm, 27 cm.

The feet are relatively still larger. He wears a shoe just a foot and a half long, while the actual total length of each foot is 14 inches, 35.5 cm., and the circumference around the instep is $11\frac{1}{2}$ inches (29.2).

There is no especial asymmetry of physique except in respect to the face, as described. The left shoulder is, however, a little higher; he is decidedly round-shouldered, and there is a slight dorsolumbar lateral curvature of the spine.



FIGURE 7. Same, showing facial hemihypertrophy.

He has no cutaneous eruptions, no pigmentation or discoloration. He has thick, coarse hair, but no beard. His muscular system is but moderately developed; the grasp of his hand is weak; he does not like to climb stairs; he has not much strength. He has good co-ordination; is a good shot. His knee-jerks are slow and feeble.

Vision is good in both eyes, and he has no contraction of the visual field. The pupils react normally.

The eyes are small, the palpetral fissure measuring 3 cm.

His intelligence is good. He sleeps well and eats well. He has a prodigious appetite, and on one occasion ate 27 plates of ice cream at one sitting, thereby winning a wager that he could eat more than two men. No unpleasant after-effects were reported. He has slight headaches at times.

His pulse beat and respiration and his heart action were normal.

I could not say whether the thyroid was changed in size. It is certainly present.

REMARKS.

The interest of this case lies first in the giant growth, and next in the progressive facial hemihypertrophy.

That gigantism is sometimes associated with acromegaly, has been shown by my own case and that of others cited.

This patient has some symptoms belonging to acromegaly, viz.: The enlargement of the bones of the left side of the face, beginning at about puberty, the kyphosis and sclerosis, the enormous feet, the coarse hair, feeble muscular development and prodigious appetite.

The progressive facial hypertrophy is very interesting on account of its rarity and its association with the gigantism.

Dr. D. W. Montgomery recently reported a case of this disease (*Medical News*, July 15, 1893,) and collected the literature of the subject.

He finds only nine cases on record, his own case and mine making eleven. In seven of them the disorder was congenital. Those which are not congenital develop at about the time of puberty or earlier. None of them occurred in persons of great size, or in acromegaly.

None of the cases resemble, in any way, those of symmetrical or irregular osseous overgrowth, and none of them were cases of leontiasis ossea. Still it seems to me that in Dr. Montgomery's case the question of leprosy might be raised. My case differs from Dr. Montgomery's, and most if not all others in the absence of involvement directly of the skin and soft tissues. Ordinary cases of unilateral facial hypertrophy have some resemblance to facial atrophy.

It is this bringing together of diseases which seem to touch each other, that I think is most instructive in giving broader ideas of pathology and enabling us to unify and simplify our conceptions of morbid processes. Hence it seems to me that the cases I present have practical interest and value.