

thus negating the presence of glycuronic acid, there seems to be no further doubt that there is exhibited in this sample of urine a well-developed case of pentosuria, as precisely the same results as above described were obtained in a specimen of urine from the same individual examined eleven months after the first report.

### ADIPOSIS DOLOROSA WITH MYXŒDEMATOUS MANIFESTATIONS.

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ADIPOSIS dolorosa is not a disease entity, but a condition characterized by a more or less definite symptom-complex. The two phenomena which are always encountered are irregular deposits of fat in the subcutaneous tissues, and pain in the fatty swellings. The other characteristics which are usually but not inevitably present are weakness, mental disturbances, and disorders of menstruation. There are other symptoms which may occasionally supervene, as sensory disturbances, headache, and diminished perspiration. It is an affection occurring in the vast majority of instances in women, generally setting in between the fortieth and fiftieth years of life, at a period when the menstrual functions are more or less deranged or the menopause has already become established. As but fifty cases of the condition have been reported in literature since Dercum first described the symptom complex in 1888, every additional instance of it should be recorded, especially if it presents certain features of myxœdema, as does the case narrated in the following:

Mrs. E. W. was referred to me by Dr. J. Gutman, of New York City, on April 21, 1908. The patient is a native of Roumania, belongs to the Hebrew race, is forty-two years old, married for eighteen years, and has never been pregnant. Her immediate family history is good; alcoholism, insanity, epilepsy, tuberculosis, or cancer had not occurred in a single member thereof. Her husband has never been affected with any venereal disease. The patient is bright, well educated, and apparently anxious to assist her medical adviser.

Menstruation started in the thirteenth year, but was never profuse. Since puberty the patient has not had an infectious disease necessitating staying in bed. About four years ago she slowly began to grow fat. At present she weighs 224 pounds. For the last year she noticed a progressive weakness, palpitation, and shortness of breath on even the slightest exertion, and, as she herself explains, "tiredness of soul and body." She has had non-

pitting edema of the face, legs, and feet, and suffers with paroxysms of pain in the entire legs, especially in the calves and the posterior portions of the thighs. The pain becomes excruciating on standing, and even more so when walking. There are also pains in the region of the neck and chest, but they are of a more fleeting nature. For the past two years the perspiration is noticeably reduced, even during hot weather, and the hair has been falling out in a marked degree.

*Examination.* The patient has an apathetic, bloated physiognomy, is well developed, and has large diffused deposits of fat over the abdomen, shoulders, and back, but desultory accumulations of fat are particularly noticeable just above the ankles, on the posterior and inner sides of the calves, the inner sides of the knees, and the external sides of the thighs and over the lower portion of the buttocks. While there is but little pain elicited on pressure upon the abdominal, cervical, and dorsal fat layers, the sensitiveness of the fat deposits in the legs is very pronounced when pressure is applied. The lower the location of the fat deposit the greater proves the pressure pain. The most acute pain is induced in the fat pad around both ankles; the pain induced in the fat deposits on the inner side of the knees and over the calves is less sharp, but much more intense than the induced sensitiveness in the fat masses of the thighs and buttocks. There are no lumpy fat deposits in the upper extremities. The circumscribed fat masses in the lower extremities are quite firm and do not pit when pressed upon. The integument directly over and surrounding the circumscribed fat deposits is soft and pliable, having retained more or less its normal character. In the face, however, the skin is rather firm and inelastic, and exhibits areas of distinct roughness. There is a slight enteroptosis, and the flabby panniculus hides the external genitals. The respiratory organs show nothing abnormal; there exists without a doubt a fatty infiltration of the heart. No cardiac lesion besides this is discernible, but the second aortic sound is distinctly accentuated. The blood pressure over the radial artery is 90 mm. Hg. by Potain's instrument. The thyroid gland is not palpable. The cervical and axillary glands are not enlarged, but just above the clavicles there are small superficial nodules which seem to be due to the shrivelling of the skin and the subjacent structures. The liver is normal in size and presents nothing unusual around its lower border. The spleen is not palpable, and its area of dullness appears to be of normal extent. The stomach is somewhat ptotic, but not enlarged. There is a marked pressure pain in the region of the descending colon and the sigmoid flexure; this is undoubtedly caused by hardened feces accumulated therein. The inguinal glands are not enlarged. The temperature, taken by mouth and rectum, is normal. Muscular power is much diminished, and the patient cannot hold the arms extended without much effort. There is greater muscular strength

in the lower extremities, but even a slight exertion causes more or less pain. The patellar reflexes and the tendo Achillis reflex are diminished. The deep reflexes of the upper extremities are sluggish. Babinski's reflex cannot be elicited. There is no tenderness over the nerve trunks. Coördination, sense of orientation, and stereognostic sense seem unimpaired. The ocular phenomena are negative. The percentage of hemoglobin is 70. There are no abnormal urinary features, neither chemical nor microscopic.

A *diagnosis* of adiposis dolorosa with symptoms of myxœdema was entertained.

The *treatment* advised consisted of an anti-obesity diet, thyroid medication, and physical therapeutics, especially vibratory massage and exercise.

May 10, 1908. The patient has executed the prescribed treatment under the supervision of her family physician. Her present weight is 210½ pounds. She feels weak, but is not discouraged. The pains in the legs continue; they are so severe in the morning that she cannot make any steps, but is compelled to crawl on her knees if she has to move about. Later in the day the pains subside to some extent, and then she is able to take some exercise. The findings of the physical examination do not materially differ from those obtained on the previous occasion. The twenty-four hours' urine amounts to 1200 c.c.; there are no pathological elements contained in it. In addition to the former treatment special exercises for the feet and a supporter for the pendulous abdomen are recommended.

July 15, 1908. The patient has religiously executed the treatment with the aid of her physician. She weighs now 188½ pounds. She complains of "general weakness from head to feet." She looks exceedingly anemic. The thyroid had to be temporarily withdrawn on account of increased cardiac palpitation and some arrhythmia. The physical examination demonstrates a stronger cardiac impulse. The radial blood pressure amounts to 120 mm. Hg. taken with Potain's instrument. The urine remains unchanged. Glucose in clinical amounts cannot be ascertained. The treatment is continued excepting the thyroid medication, which is to be resumed in a week or two, but in smaller daily doses.

January 4, 1909. The patient presented herself today to show me the beneficial effects of the treatment. Excepting the pallor, which, she says, has always caused her much annoyance since her early youth, she looks very well. She feels strong, and is able to walk from five to eight miles a day; she experiences no shortness of breath on ordinary exercise, but perspires mildly when she walks briskly. The fat bunches have disappeared almost entirely; the neuralgic pains ceased about four months ago; there is no tenderness on pressure on the location of the former fat masses. The skin in the supraclavicular regions and in the face has been quite

tender. She evinces not the slightest mental depression and apathy, but, on the contrary, displays a healthy optimism. Her present weight is 161 pounds.

COMMENTS. The foregoing case is of especial interest for two reasons—the concurrence of adiposis dolorosa with well-marked myxœdematous manifestations, and the synchronous abatement of the symptoms of both affections after thyroid medication and a general anti-obesity treatment.

Symptoms of myxœdema are known to supervene in a certain proportion of the cases of adiposis dolorosa. Dereum's first publication on this subject was entitled "a subcutaneous connective-tissue dystrophy resembling myxœdema," and Henry reported some time later a case under the caption of "myxœdematoid dystrophy." Thus, the possible concurrence of both conditions, or at least a certain resemblance of adiposis dolorosa with myxœdema, was recognized by the early writers on the syndrome.

The instance of adiposis dolorosa under consideration exhibited the following myxœdematous manifestations: Apathetic, bloated physiognomy; history of non-pitting œdema of face, legs, and feet; firm, inelastic, and rough skin in the face (seat of former œdemas); falling out of the hair; non-palpable thyroid, and dermal tumefactions in the supraciliary areas. This symptom complex alone would prompt the diagnosis of myxœdema, but it was overshadowed, at least for the time being, by the acuteness of that of adiposis dolorosa. The association of the two syndromes is by no means accidental, although it appears that in none of the cases of adiposis dolorosa on record were the myxœdematous features as plentiful and well pronounced as in the case before us.

In view of the frequency of myxœdematous symptoms in adiposis dolorosa, I think we are justified in accepting a kindred cause of both syndromes. That thyroid insufficiency stands at the foundation of myxœdema there can be little doubt; again, some thyroid alteration was found in four out of five cases of adiposis dolorosa which came to autopsy. While the seat of the externally visible pathognomonic symptoms of myxœdema is in the subcutaneous tissues, that of adiposis dolorosa is situated in the fatty structures.

Moreover, the improvement of my case following the administration of thyroid extract seems to evince with certainty that perverse thyroid function was, so say the least, an antecedent. The yielding of both symptom complexes to the same medication again points to their interrelation or their springing from a kindred cause. Thyroid therapy cannot, therefore, be utilized as a test of differentiation between myxœdema and adiposis dolorosa, as some authors maintain, because both syndromes may vanish under its influence, and, as in the present instance, even at the same time. In so far as the pains disappeared in the ratio of shrinkage of the fat bunches, we are justified in concluding that the irritation of the nerve terminals

was either due to mechanical insults on the part of the overgrowth of fat tissue, or to certain fatty acids or products of katabolism exciting the nerve trunks in the vicinity of the fat deposits and stimulating the fat-tissue to further proliferation. In view of the fact that myxœdema occurs without pains in the swellings, it appears that thyroid insufficiency cannot be held directly responsible for the aches and paroxysms in adiposis dolorosa.

## HEMOPHILIA WITH THE REPORT OF A CASE OF TYPHOID FEVER IN A HEMOPHILIC SUBJECT.

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By the term hemophilia we mean a peculiar hereditary anomaly of constitution characterized by traumatic hemorrhages of great stubbornness and also by a conspicuous tendency to spontaneous and repeatedly recurring hemorrhages, subcutaneous, subfascial, intra-articular, intraperitoneal, intrapleural, intracranial, or from the mucous surfaces. The condition, of course, must not be confounded with other pathological states, such as scurvy and purpura, in which there is a marked tendency to hemorrhage; the former is a definite disturbance of nutrition, the latter an acquired condition dependent upon a variety of specific organisms or their toxins. Nor must we confound it with the so-called hemorrhagic diathesis associated at times with certain of the specific fevers, nor with multiple hereditary telangiectasis first described by Osler.<sup>1</sup>

According to Wilson,<sup>2</sup> priority is due Albucasis, an Arab, who died in Cordova A.D. 1107, and in whose writings a well authenticated instance of hemophilia is mentioned. Alexander Benedictus, in 1539, Hochsetter, in 1674, Legg, in the same year, Fordyce, in 1784, and Rave, in 1798, were among the earlier observers. The first American physician to mention the disease was Otto,<sup>3</sup> in 1803, and it was he who applied the term "bleeders" to patients afflicted in this way. He gave the history of a family in which the disease could be traced back 80 years. In 1813 Hay gave details of the Appleton-Swain family, which, it seems, was another branch of the family reported by Otto. In 1817 Buel<sup>4</sup> reported cases in the Collins family, and in 1828 Coates<sup>5</sup> recorded others in a Pennsylvania family. The German school here took up the study, and articles appeared by Nassi, Wachsmuth, Virchow, and

<sup>1</sup> Johns Hopkins Hosp. Bull., November, 1901.

<sup>2</sup> Practitioner, Lond., 1905, lxxv, 829.

<sup>3</sup> Trans. Med. and Phys. Soc., New York, 1817.

<sup>4</sup> North Amer. Med. Jour., 1826.

<sup>5</sup> New York Med. Repository, 1803.