A CLINICAL LECTURE ON A CASE OF HYPERTROPHIC PULMONARY OSTEO-ARTHROPATHY.

Delivered to the Students of the Melbourne Hospital.

By J. W. SPRINGTHORPE, M.A., M.D.MELB., M.R.C.P.,
Physician to the Melbourne Hospital, Lecturer on Therapeutics in the
Melbourne University.

E. S., aged 21, was admitted into the Melbourne Hospital on January 23rd, 1893. His father was gouty, and died after an injury at 46. His mother is alive, neurotic, and suffering from climacteric neuroses. Of the 13 children 4 are living, 2 being slightly delicate, and 1 suffering from (?) hysterical paraplegia. Three died in infancy, 1 from marasmus and, later in life, 5 died from marasmus, abscess in the lung, typhoid fever, diphtheria, and burns respectively.

The patient himself was always strong and well up till 10

years of age. Neither mother nor elder sister noticed any abnormality of skin or bone, and none is visible in a photograph taken about the age of 8. At 10 he had an attack of right pleurisy with effusion, which was said to have been "followed by typhoid fever." He was very seriously ill for three months, and the probability seems that an empyema was then in progress. He continued ailing, and some three months later began to exhibit some curvature of the spine, for which he was put in a plaster-of-paris jacket at the Children's Hospital. Whilst there he was treated also for a skin affection, about which no notes can be found (the notes generally having been mislaid), but his then attendant, Dr. Snowball's, strong impression is that it was ichthyosis. Nothing abnormal was noticed about the joints or bones. The abnormalities in spine, skin, and bones increased gradually to their present condition. For the past two years there has been transient swelling of the feet, and for the past six months swelling of the stomach.

On admission the patient complained of dyspnoea, pain and swelling in the legs and stomach, weakness, and cough; the face also was swollen; the urine 1010, in fair quantity, and free from albumen; the temperature 97.4°; the right chest was very much flattened and retracted; the liver enormously enlarged; a medium quantity of fluid in the abdomen; the heart considerably enlarged, with thrill, heaving diffused impulse, and murmurs both systolic and diastolic over mitral and aortic areas. In addition he presented the following uncommon combination of signs in spine, bones, joints, and skin:

1. There was a very marked curvature of the spine, occupying the whole of the dorsal region; behind it was on a line with the end of the spine of the left scapula. The right chest in the intermammary line measured $5\frac{1}{2}$ inches less than the left, the measurements being $11\frac{1}{2}$ and 17 inches respectively. The sternum was displaced upwards and to the left, the left ribs higher than the right, and the head carried well over to the right side.

2. In addition to general wasting, the pectorals, the muscles of the scapulae and upper arms were reduced almost to nothing. The skin over the abdomen, arms, forearms, and hands was thick and scaly; it was even more thick and scaly on all the surfaces of the lower extremities, especially the lower front of the thighs, knees, and the front of the ankles, where it could not be described as anything but ichthyotic. The colour was livid grey, except about the knees and ankles, where it was dirt-ingrained. The penis was that of a boy of 10 or thereabouts, the scrotum large, the cremasteric reflexes present, but there was no history of any erotic excitation at any time. Beneath the thickened epidermis of the legs and thighs was some cedema.

3. There was also noticeable enlargement,

with deformity of the extremities, with, apparently general lengthening of the long bones. On each side the measurements were:

From tip of acromion to olecranon 12 in.; from olecranon to lower end of ulna, 10 in.; from carpus to end of second finger, 7 in.; from trochanter to external condyle 16 in.; from condyle to external malleolus, 16 in.; from heel to end of big toe, 10 in.

The lower ends of both radius and ulna were enlarged and prominent, measuring $6\frac{1}{2}$ inches round the left and $6\frac{1}{2}$ inches on the right side. Both hands were enlarged, being increased in length but not in

breadth; the fingers were lengthened, especially at the ends, and very markedly clubbed. There was also some prominence of the knees, the circumference being $13\frac{1}{2}$ inches, and unusual prominence and thickening round the ankles, and both big toes were much enlarged. Unfortunately, his unexpected death a few days after admission prevented further and more extended examination.

4. The face presented no abnormalities.

Post-mortem Examination.—It was, unfortunately, only after considerable difficulty and delay that a limited *post-mortem* examination was obtained. The examination, conducted by our assistant pathologist (Dr. Mollison), disclosed the following: Abdomen distended, greenish, containing a moderate amount of fluid. Liver pushed down almost to anterior superior spine. No beading of ribs. Pericardium everywhere adherent. Heart large, flabby and pale; cavities much dilated and containing fluid blood; no valvular lesion and no congenital deformity; no thymus noticed; no enlargement of clavicles. Right pleural cavity—one large abscess cavity—still containing some pus; its walls irregular, thickened, pigmented, and in patches calcareous. The right lung com-

Hypertrophic pulmonary osteo-arthropathy; side view. Photograph taken after death by the coroner's orderly, Constable Davidson.

pletely collapsed; no connection between the empyema and spine; no necrosis of bone. The left lung rather small, engorged, and friable; no enlargement of glands or blood vessels. Liver, weighing 5 lbs. 6 ozs. (? lardaceous), decomposing. Spleen 12½ ozs.; fibroid element increased (? lardaceous). Kidneys pale, decomposing. Brain: pituitary body not enlarged; no increase in the fossæ at the base.

REMARKS.—The foregoing case is, I believe, the first of its kind reported in Australia. As with Friedreich's first cases, it was some years ago taken for a case of osteitis deformans, and at first sight regarded by myself as acromegaly. Further examination, however, especially in the light of Dr. Souza-Leite's monograph on *Acromegaly* (New Sydenham Society, 1891) showed that it was rather one of what Marie has called "Ostéo-arthropathie hypertrophiante pneumique," or hypertrophic pulmonary osteo-arthropathy.

The disease is a rare one, and only a few cases have as yet been recorded. Marie¹ has reported 8 cases, 2 of Friedreich-Erb,² and single cases by Sollier, Lewis, Saundby,³ Fräntzel,⁴ Ewald,⁵ and himself. In May, 1890, another case was reported by Spillmann and Haushalter,⁶ and in December, 1890, a tenth by Gerhardt.⁷ During 1891 another was reported by Rausier,⁸ whilst Julius Arnold⁹ and Bamberger¹⁰ have discussed respectively the diagnosis and pathology. I can find no reference to any other cases in the *Index Medicus*, *D. M. W.*, *Sajous's Annual*, the *Lancet*, or the *BRITISH MEDICAL JOURNAL* for either 1891 or 1892. The present thus seems to be the twelfth recorded case.

DIFFERENTIAL DIAGNOSIS.

In the present state of our knowledge, we can readily exclude osteitis deformans. In osteitis deformans the principal symptoms described are as follows: Considerable increase in the size and curve of the long bones and of the bones of the head and trunk, producing a characteristic appearance, the femurs and tibiae being curved forwards, the legs abducted, and the neck and trunk fixed as in rickets in a forward flexure, respiration impeded and diaphragmatic from the hypertrophy and ankylosis of the ribs, a history of lightning pains, a beginning in mid age, and protracted duration without marked affection of the general health.

Symptoms.	Acromegaly.	Hypertrophic Pulmonary Osteo-arthropathy.
Upper Extremity	Hands "spade-like," enormously widened and thickened, though not lengthened; fingers "sausage-shaped," thenar and hypothenar eminences enlarged, nails small, wrist in proportion; as a rule, the hypertrophy ceasing at wrist	Enormous and deformed; not enlarged in carpo-metacarpal region, but in fingers, especially the last phalanx; nails enlarged and curved; lower ends of radius and ulna forming an enormous projection above the hand.
Feet	Similarly enlarged, thickened, flattened; hypertrophy ceasing at ankle	Like the hands; last phalanx enlarged; the ankle thickened and deformed, the tarso-metatarsus least hypertrophied.
Head and Face	Face enlarged, elongated, oval; lips thick; tongue hypertrophied; nose large and flat; orbital ridges projecting; prognathous lower jaw; ears large; skin pigmented	Not enlarged or oval; lower jaw not prognathous; lips, tongue, neck, and larynx not enlarged.
Spine	Marked cervico-dorsal curvature, with slight lumbar lordosis	Curvature, lower dorsal and lumbar.
General	Hypertrophy of all the tissues in the region <i>en masse</i> . Alteration in voice; headache, pains, thirst; boulimia, changes in vision	Hypertrophy of long bones, especially at articular ends. Absent.
Post mortem	Skull sinuses dilated; fossæ at the base, especially pituitary, enlarged; pituitary body hypertrophied, also ganglia of sympathetic; thymus persistent; thyroid altered, also organs of speech and sexuality; heart and blood vessels hypertrophied	Respiratory affections—for example, empyema in two, neoplasm of lung in two, and bronchitic affection in three; heart affections also.

This is a very different clinical picture from that presented by the present case. From acromegaly, however (itself a rare disease, of which the total number of cases yet reported does not amount to fifty, and of which no instance has as yet, so far as I know, been reported in Australia) the diagnosis is a matter of greater difficulty. Still the accompanying table arranged from Marie's paper, as quoted in Souza-Leite's monograph, will make the distinction evident.

The case is thus, without doubt, one of hypertrophic pulmonary osteo-arthropathy. Its identity is still further illustrated by comparing the photograph with the woodcuts figured 20, 23, and 24 in Souza-Leite's thesis. The explanation of this peculiar disease is still only problematical. Marie, accepting gout and rheumatic affections as analogies, attributes it to the effect on the osteo-fibrous structures of the toxins of septic organisms, which have been absorbed into the general circulation from lesions in the respiratory tract. Bamberger finds himself unable to offer a solution which will cover all cases, and considers it probable that several causes exist favouring the theory that some chemical substance like phosphorus is produced in the diseased lungs, and acts as an irritant to bone.

In the present case the sequence of events seems to have been somewhat as follows: A family history with neurotic and tuberculous tendencies, an unrecognised empyema with adherent pericardium, extreme and permanent collapse of the lung with consecutive dorsal curvature, finally nutrition and development carried on under the double disadvantage of serious heart disease and much impaired oxygenation of the blood, with abnormalities present in the epidermis, and certain parts of the bony system. Several interesting problems present themselves:

1. The exact nature of the osteo-arthropathy. Bamberger calls this a periostitis. In my case no further examination was permitted. My only other note is that there was no other crepitus in the joints. Again, why are wrist and ankle, and also knee affected, whilst shoulder, hip, and elbow are apparently unaffected? Whatever be the true explanation it is suggestive to find that it is the lower articular ends of the long bones which are distinctly increased in size, and, so far as my remembrances in anatomy serve me, these seem to be the parts in which the greatest amount of developmental activity would be then taking place.

2. The kyphosis in this case occurred early, and apparently as a result of the collapse of the lung, and not as in some of Marie's four cases with curvature, some years after the other bony deformities, and dependent on lesion of the intervertebral articulations.

3. The condition of the skin is somewhat puzzling. It could scarcely be described as anything else than ichthyosis. The complication is absent from Marie's description, and is probably an accidental accompaniment of the present case. Though no history of such affection was procurable in the family, or remembrance of its presence in early life, still there is evidence of its presence when being treated for curvature at the age of 10. In all probability it existed still earlier in a mild form, and gradually reached its later noticeable manifestations.

4. The clubbing of the fingers was exceedingly marked, and exhibited in an extreme degree what may occur in ordinary chronic heart and lung disease. The other *post-mortem* appearances speak for themselves.

Theory as to Causation.—Dealing simply with the case under notice, the circumstances and sequence of events suggest in my mind a probable causation which I would advance only with great diffidence, and for purposes of further investigation. It is that this rare manifestation of disease may be the result of exceptional abnormality of development under the influence of continued and uncommon maloxxygenation of the blood. Such a hypothesis seems to explain, to some extent at least, why certain portions of the long bones are

¹ *Revue de Médecine*, January, 1890, and *Bulletin Médical*, December, 1890.

² *Deutsches Archiv für klinische Medizin*, 1888, Bd. 42, p. 304.

³ *Illustrated Medical News*, March, 1889.

⁴ *D. M. W.*, August, 1888.

⁵ *B. K. W.*, March, 1889.

⁶ *Rev. de Méd.*

⁷ *B. K. W.*

⁸ *Rev. de Méd.*, p. 30.

⁹ *Ziegler's Beiträge*, 1891, B. x, p. 1.

¹⁰ *Zeitschrift für klinische Med.*, 1891, B. xviii, p. 193.

specially affected. It already explains the extreme clubbing of the fingers; and if we, following Erasmus Wilson, regard ichthyosis as "a defect of development and normal growth of the skin," it explains also why in the present case the skin took such a prominent part in the abnormalities produced. How far such a hypothesis fits in with the requirements of the other recorded cases I am, in the absence of information thereon, quite unable to express any opinion.

NOTES OF TWO CASES OF ACROMEGALY.

By W. B. RANSOM, M.A., M.D., M.R.C.P.,
Physician to the General Hospital, Nottingham.

THE following are two typical cases of this interesting disease:

CASE I.—Miss M., aged 47, is of Irish birth, but has lived for ten years in New York, from whence she returned to England in the summer of 1894. She says that as a young woman she was slight and active, and had rather small hands and feet. For the last five or six years she has noticed enlargement of the hands, feet, and face, though she is not clear as to which enlarged first. It is five years since she last menstruated. During that time she has grown nervous, irritable, and languid, and thinks her memory is failing. Her vision has become very defective. For a year or two she has perspired very freely, and has been intolerant of heat, though she stands cold fairly. She has had no loss of hair, hardly any pains in the limbs, and but slight headache. She says that an American doctor whom she consulted a year ago told her she had myxedema, but that another doctor said she had not. She had taken thyroid tablets for some time without benefit. There is no history of any similar disease occurring in any member of her family.

Fig. 1.—Case I.

When I first saw her, in August, 1894, the patient presented the characteristic countenance of acromegaly. The nose was long and thickened, the lower jaw large and prognathous, the lower lip swollen and everted, and the skin of the face generally coarse, thickened, and wrinkled. The

eyebrows were of average quantity, and the hair of the head was not markedly altered. (In the photograph (Fig. 1) the hair has been touched up by the photographer who printed from my negative.) The hands were much enlarged, apparently both in the hard and soft parts, the fingers being uniformly enlarged, without bulbous extremities, and with flat square nails. The wrist, relatively to the metacarpus, was small. The feet were also increased in size. Owing to the nervous and almost hysterical state of the patient, a proper examination of the chest and abdomen was not feasible, but no kyphosis was obvious when she stood up in her clothes. The tongue was extremely large. The thyroid appeared of normal size, and there was no unusual dullness behind the manubrium sterni. The voice was somewhat "croaky" in character.

The patient complained of considerable shortness of breath on exertion, but the pulse was of fair quality, and there was no sign of arterial degeneration. The apex beat of the heart was just outside the nipple, but otherwise the organ presented no abnormal sign. The lungs were normal. There were varicose veins in the left leg. The patient slept fairly, was troubled only with slight headaches and slight occasional "rheumatic" pains in the limbs. The digestive system appeared normal, as was the urine. Vision was markedly impaired, and she could only read large print with strong convex glasses. Perimetric examination showed bitemporal hemianopia (Fig. 2). The pupils were normal, and there was

Fig. 2.—Visual fields of Case I.

no ocular or other paresis. The optic discs were rather small, pale, and irregular in outline, but the fundi were otherwise normal. The patient was treated with arsenic and tabloids of pituitary gland, but she grew gradually weaker and more languid in body and more fretful in mind. In February of this year she succumbed to an attack of bronchitis. No *post-mortem* examination was allowed.

CASE II.—Mrs. B., aged 34, came to the hospital in October, 1894, for general lassitude and excessive thirst. She first noticed enlargement of the hands about five years ago. At that time she had an illness called influenza, in which she had rigors, fever, and frontal headache, being ill a fortnight. This illness left her weak for some time, and a few months later she began to have severe headaches in the forehead and at the root of the nose, perhaps as often as twice a week. The hands continued to increase in size, and two years ago the feet did so also, so that instead of "fours" in boots she had to wear "sevens." The face and head began to grow large about the same time, and she says that two years ago her sight suddenly became dim, and she only saw one half of an object she was looking at. She thinks she increased in height as she certainly did in weight, rising from nine to thirteen stones. During the last eight months, however, she has been very thirsty and hungry, has passed a great quantity of urine, and has lost three stones in weight. Her strength has been much diminished during this time.

Whereas two years ago she began to sweat profusely, for the last eight months she has hardly sweated at all. The headaches have remained unaltered during the last five years. She has three children, aged 12, 10, and 8, and she has not menstruated since the birth of the last. The menstrual function has been always scanty; from the age of 14 to that of 22, when she married, she suffered from symptoms of anæmia, and menstruation was very infrequent, while since marriage it has only occurred once or twice.