

ADDITIONAL  
CASES OF OSTEITIS DEFORMANS.

BY

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I SHOULD not have offered to the Royal Medical and Chirurgical Society a mere collection of cases such as this paper contains, if it were not for the hope that they may help to clearly indicate the chief characters of the disease to which I venture to give the name of osteitis deformans, and which, so far as I know, was first described in the paper published in the 60th volume of the Society's 'Transactions.'<sup>1</sup> Since that time, about five years ago,

<sup>1</sup> After the publication of the paper I found that the name *ostitis deformans* had been given by Prof. Czerny, of Freiburg, to a disease described by him in the 'Wiener medizinische Wochenschrift,' September 27th, 1873. It is mainly, as he says, "Eine lokale Malacie des Unterschenkels," a rather acute inflammation of the lower part of the tibia and fibula, inducing softening and angular bending, and then followed by hardening. A specimen of the only instance of this disease that I have seen has lately been presented to the Museum of the College of Surgeons by Dr. Butt, of Hereford. Like all Prof. Czerny's cases it occurred in a young man.

It may be well to add also that the sternum, a clavicle, and a rib of the man, whose case by Saucerotte is referred to in my last paper, are in the Musée Dupuytren. They were obtained by Saucerotte many years after his account of the case was published. The description of them in the 'Dupuytren Catalogue,' vol. ii, p. 148, leaves it very doubtful whether the disease was osteitis deformans.

I have seen seven cases of the disease, and have recorded concerning them the facts which follow :

CASE 1.—May 29th, 1878. A lady, æt. 65, having no appearance of general ill-health and looking her age, complained chiefly of what she deemed to be rheumatic and neuralgic pains of her back and lower limbs. She ascribed them to exposure to cold thirteen years ago ; for she had rarely since that time been free from pain, and had lost strength and health ; and in the last year or more, had suffered with what she considered to be attacks of bronchitis and asthma.

Soon after the beginning of her pain, that is, about ten or twelve years ago, her daughters thought that she was losing in height, and that the shape of her head was changing ; and from that time she had been becoming less tall, till now she had lost four inches and a half in height, and stooped low with her head forward and her chin raised.

She had marks of slight gouty affections in some knotted knuckles, and frequent flatulence and occasional excess of lithic acid in the urine. But she had never had fever, ague, or any acute illness, and had borne five children in rather hard labours, without ill consequences.

Her father had been gouty and died in old age ; her mother died young after parturition. She did not know of any case of scrofula, consumption, or cancer having occurred in her family.

Her head, though she said it had always been a remarkably large one, was certainly enlarged, and chiefly by convex bossed additions over and about the junction of the frontal and sagittal sutures and above the temporal sutures. They were symmetrical, and might be guessed to be additions of one third or half an inch in thickness, perhaps additions to a general thickening of the cranium. But they neither were nor had been associated with headache or any other local trouble, and sight, hearing, and the other senses were unimpaired.

The dorsal spine was curved with a very marked posterior rounded curve, inclining a little to the right, without any compensating curve to the left below it. The curve produced a low stooping posture, with very prominent right shoulder, and might be estimated as shortening the trunk about two or two and a half inches.

The ribs were nearly horizontal, flattened at the sides, and, even in deep inspirations, nearly motionless. The respirations were almost wholly diaphragmatic, with elevation of the sternum. They appeared to be sufficient during quietude, but in any hurry or mental emotion, or any unusual exertion, great distress of breathing was felt, and walking upstairs seemed even dangerous; she was always carried up.

The lumbar spine appeared of natural form, so did the pelvis, and likewise all parts of the upper extremities.

The femora were exceedingly curved outwards and forwards; the left rather more than the right. Their shafts felt in their whole length, especially, I think, in their lower half, large, rounded, thickened.

Similarly the tibiæ were curved forwards and were very large in their whole length. Their anterior surfaces felt nearly twice as wide as in nature, smooth, and with large rounded margins.

The feet and all the articulations of the lower limbs appeared quite healthy. None of the enlarged bones were tender on pressure.

The likeness of the forms of the trunk and lower limbs in this case, and in the first case recorded in my former paper, was very striking. The similarity of disease could not be doubted.

The patient lived two years and a half after this note of her case was made, and during this time was under the care of Mr. Haynes, of Stansted, to whom I am indebted for being able to report that little change ensued in the bones of the lower extremities; that the skull became more deformed, especially with a broad, high boss along its upper middle line; and the spine more curved

and prominent in its dorsal part. Death ensued in consequence of Bright's disease and valvular disease of the heart, with extreme anasarca. It did not appear due in any degree to the disease of the bones, unless it were that the difficulty of breathing was aggravated by the deformity of the chest. There was no indication of cancerous disease of any part.

Examination after death was not allowed.

CASE 2.—February 17th, 1879. A man, æt. 62, looking in everything but shape completely healthy, and feeling well and fit for work, even more fit than ever for any mental work, told that he had been healthy all his life, and had been six feet and a half inch high, slim, well made, and active, a volunteer, and given to all kinds of exercise. He was not aware of any unhealthy inheritance, certainly not of gout. He did not know the very beginning of his illness, or any cause for it. He first observed, distinctly, that his stature was less, and that his left limb was rather stiff and lame, in May, 1871, when he was 54. A bone-setter then told him that his hip was "out" and professed to reduce it, and later in the year he was treated for chronic rheumatism at Buxton and obtained there some relief from pain. From this time he appeared to be constantly losing height and becoming more misshapen; but he had never suffered more than might be ascribed to chronic rheumatism of moderate severity in the spine and limbs, and had never left off work or the taking of moderate exercise. He had been obliged to wear larger hats than he had been accustomed to, and had twice or more in the last two years increased the size, but he never had a headache of any kind. Once only in the eight years he had been ill. This was with jaundice, of no great severity, in 1876, and once or twice he had had dyspepsia. Lately he had become rather deaf.

Now he was five feet eight inches, having lost four inches and a half in the eight years. His head was large and well formed, but disproportionate to his face, and his

frontal veins looked very full and tortuous. The face was natural. His neck was very short and stiff, and the head was habitually held forward with the chin rather in advance of the sternum, and not more than two inches above it when the face was directed straight forward. The spine was very short, nearly straight, as if its anterior and posterior curves were straightened in the shortening, and it was far too little flexible. The ribs were crowded, nearly horizontal, and hardly raised in inspiration. His in-breathing was almost wholly with the diaphragm, and, when deep, with great uplifting of the shoulders. Yet he called it "pleasant breathing," and was never troubled with it unless on fast walking. The abdomen was short, prominent, overhanging, deeply folded. The clavicles were very big and full-curved, twice as big (at a guess) as they should have been. The arms and hands appeared quite natural, muscular, and agile, with all the power of writing, fencing, and other uses that they had ever had. I observed nothing wrong in the pelvis. The femora were big and very curved; the left, which had been longer and more affected than the right, was curved outwards and forwards, the right forwards and but little outwards. The tibiæ were big in their whole length, broadly rounded, very curved forward. The feet and all the joints of the lower limbs appeared quite healthy, but the mobility of the hips and knees was lessened.

This patient still lives in fair general health, but with all his deformities increased.

CASE 3.—April 12th, 1879. Mr. M—, æt. 66, formerly a colonial judge, looking older than his years, and dry and rather shrivelled, had had generally good health, but once had fever, and once, about fourteen years ago, congestion of the brain. He had been liable to general rheumatic pains, but not more in the lower limbs than in other parts, and he came for advice about his left ankle, which had become less movable in consequence of the

great overhanging of the tibia and the very acute angle in which he habitually held it whilst standing.

His appearance and posture were very characteristic—with his head forward, his shoulders raised, short trunk, legs apart, feet turned out—the right foot forward, the left behind—and both legs greatly curved. Except that he was a shorter and smaller man, the photographs published in the 60th volume of the 'Transactions' might have suited him.

In ten years his height had diminished about two inches, chiefly, I think, from shortening of the spine. The head was well formed and large—it always had been so—and he had always had difficulty in finding a hat large enough for him; but the only indication of increase in it which he had observed was that his present hat was rather tight. The enlargement, however, if any, must have been very little. I could not find any sign of enlargement in the upper limbs. The ribs were nearly horizontal, and less mobile than they should have been. There was less than a finger's breadth between the lowest rib and the crest of the ilium. The spine was not curved below the lower part of the neck. Each femur in its lower and middle thirds felt thick, and curved forwards and outwards, the right more than the left; the necks of the femora were lowered. The tibiæ were in their whole length very big, rounded at their edges, and exceedingly curved forwards; they felt uneven and knotted, and the integuments in front of them were over-hot; they overhung the feet, and in standing the legs and feet were at acute angles.

The date of the beginning of these changes was uncertain. They had, probably, been in very slow progress for not less than ten years, but they had given so little trouble that they had not been closely watched. No cause of them could be told. One brother was gouty, but the patient himself had not had any marked gout, and had lived a simple temperate life. The disease probably began in Melbourne.

CASE 4.—1880. Mr. S— was 51 years old, a very

small man, who had never weighed so much as eight stone, and now was about seven stone eight pounds.

He had so exactly the characteristic form that one could instantly feel sure of the diagnosis of his case. His head was inclined forwards and downwards, his neck looked short, his back was curved backwards, his chest low and small; his pelvis looked broad and womanly; his arms hung low. The lower limbs were arched and divergent: the right advanced, the left as if dropping back, the left foot even more than the right, flat and overhung. The head was, he believed, quite unchanged in size and shape; it was long and flat-sided, but he had not had occasion to change his hat. Both clavicles were enlarged, they felt thick, clumsy, rough. The ribs were very little mobile; the chest was lifted as in one piece when he drew his breath deeply, raising his shoulders. The chest was "square," the ribs flattened at the sides, and bending round with an angle to form the similarly flattened chest front. The ribs severally did not feel as if enlarged. The liver appeared to be pressed high up into the chest; there was dulness to the nipple. The abdomen was wrinkled and undulated as if its walls were too long. The spine was deeply and roundly curved in its dorsal part, but straight in its lumbar. The femora and tibiæ were very large, especially the lower halves of both, and all were very curved; the femora forwards and outwards, the tibiæ more forwards. The joints felt and looked healthy, but the left knee-joint was painful in movement and rather stiff. Except that he was very thin and dry, all the structures except the bones appeared quite healthy. He could find nothing for complaint but the pain in his knee, which had now lasted some weeks.

He ascribed his illness to a walk down Vesuvius ten years ago. He was very tired with it, was laid up with pains in his lower limbs, and from that time, as he believed, his legs began to curve. Since that time he had not had any considerable illness, but from time to time pains of no great severity confined to the lower limbs. Many years

ago he had had syphilis, and there were two scars of tertiary sores on the lower limbs.

I advised him to take iodide of potassium, but it did no good. He remains in good general health and with very slowly progressive changes in his limbs.

In the fifth case, which so far as the notes described, resembled the preceding, Mr. Butlin examined one of the tibiæ post mortem. The condition differed in no degree from that of the bones described in the 60th volume of our 'Transactions.'

CASE 5.—January, 1882. I have so few notes of this next case that I should not have inserted it but for the examination of a portion of one of the diseased tibiæ which Mr. Butlin was so good as to make.

The Rev. Mr. G— had been an active, healthy man, till he was nearly sixty, and he died at sixty-six; his chief signs of illness during this interval having indicated various attacks of irregular gout with bronchitis, emphysema, and diseased heart-valves. He was born of long-lived parents, and had no sign of general disease but such as might be ascribed to gout.

Indications of osteitis deformans had appeared about four years before his death and had slowly increased. The tibiæ especially had gradually become curved forwards, large, rounded, and uneven, and at first had been painful and hot. The femora were also slightly affected, and the spine had become roundly arched and over-hanging at its upper part.

Mr. Butlin reported that the portion of tibia which he minutely examined did not differ in any respect, so far as he could see, from the diseased bones of which his descriptions are recorded in the first case related in my former paper (see vol. lx., p. 46).

CASE 6.—March 9th, 1882. Mrs. S—, æt. 62, mother of three children, and generally healthy, though with inheritance of gout, and having often flatulence and various

gouty symptoms. She was very thin, and her large bossed head, low stooping, and oblique posture, and separated lower limbs, with one far in advance of the other, might at once have sufficed for diagnosis. She believed that her disease began at least ten years ago, when first she observed some enlargement of the external angular processes of the frontal bone. From that time she had had various pains regarded as rheumatic or gouty in her back and lower limbs, and had observed her head growing larger, her spine curving, her height diminishing, her lower limbs bending and becoming weaker, till now she was nearly four inches shorter than she used to be, could hardly stand without support, and walked slowly and as if her right leg was too long.

Her head appeared enlarged at every part, and measured twenty-four inches in its largest circumference; it had a square look, and was bossed, chiefly over the frontal and parietal protuberances and over the lower part of the occiput. The orbital arches also appeared large and prominent, but in the bones of the face there was no apparent change. The dorsal part of the spine was very curved backwards and to the right, the chest was shortened, the ribs crowded but fairly movable. The pelvis seemed not changed. The clavicles were very large and curved, like those of a strong man, contrasting strangely with the thin weak look of the upper limbs, whose bones were slim and naturally straight.

Both femora and both tibiæ were very curved forwards and outwards, and felt large and rugged. Especially the lower parts of the tibiæ were enlarged, and there was little diminution of size between the shafts and the articular ends, which seemed as if over-hanging the ankles. Both shins felt hot.

The bones of both hands and feet were healthy unless for some nodular enlargement of the phalanges such as one sees due to gout.

The patient was not aware of any inherited liability except that of gout, or of anything that could have induced

the disease of bones. She had never been dangerously ill.

Lately her sight had become much impaired and still more lately she had been becoming deaf.

CASE 7.—May, 1882. In this case, a lady, æt. 58, presented the usual characters of the disease in a well-marked form, but had observed the first signs of it when she was only 28. At that time, during the fatigue of long nursing a relative, she had pains in her lower limbs and found her tibiæ becoming large and curved. Her spine also at this time began to curve. The disease was very slowly and almost painlessly progressive; its chief or only seats being the tibiæ, femora, spine, and clavicles; but when she was 53 she observed enlargement and slight curvature in the lower thirds of both radii. She ascribed this to the exertions she used in frequently lifting her invalid husband, and the morbid changes had continued slowly increasing ever since. Her head was very large, measuring twenty-four inches and a half in its greatest circumference, and looking as if unnaturally prominent in the middle and upper frontal part, but she was sure that this was only its natural size and shape.

With the exception of the disease of the bones and the consequent great difficulty of walking, this lady seemed in excellent health. She had lived very actively; had had two children without any ill consequences, and knew of no inherited tendency to disease unless it were to gout. She had had various treatment with both baths and medicines, but none had seemed useful even for a time.

The seven cases now related seem sufficient when added to the five recorded in the 60th volume, to justify the giving of a distinctive name and a definite general description of the disease observed in them. It usually affects many bones, most frequently the long bones of the lower extremities, the clavicles, and the vault of the skull. The affected bones become enlarged and heavy, but with

such weakening of their structure that those which have to carry weight or to bear much muscular traction become unnaturally curved and misshapen. The disease is very slowly progressive, and is felt only in pain, like that of rheumatism or neuralgia, in the affected limbs, and in increased heat at the tibix. But neither the pain nor the heat are constant, nor do they continue during the whole progress of the disease; and pain has not been observed in the head even in the cases in which the skull was very thickened. There is not any clear evidence of general disturbance of health. In all the cases traced to the end of life, death has ensued through some coincident, not evidently associated, disease, which has been aggravated by the condition of the bones only in so far as they may have diminished the range of breathing and the general muscular activity.

At present, with the exception of the seventh case, this disease has been observed as beginning only in persons over forty years old, and it has appeared in no usual relation, whether by inheritance or coincidence, with any other disease except gout. I have not found cases to be added to those mentioned in my last paper in which it was associated with cancer.

In all the cases I have seen, the general appearance, postures, and movements of the patients have been so alike that these alone might often suffice for diagnosis of the disease. The most characteristic are the loss of height indicated by the low position of the hands when the arms are hanging down; the low stooping, with very round shoulders and the head far forwards, and with the chin raised as if to clear the upper edge of the sternum; the chest sunken towards the pelvis, the abdomen pendulous; the curved lower limbs, held apart and usually with one advanced in front of the other, and both with knees slightly bent; the ankles overhung by the legs, and the toes turned out. The enlarged cranium, square looking or bossed, may add distinctiveness to these characters, and they are completed in the slow and awkward gait of

the patients, and in the shallow costal breathing, compensated by wide movements of the diaphragm and abdominal wall, and in deep breathing by the uplifted shoulders.

I have seen no case in which these characters are imitated except those of ankylosis of the vertebræ and ribs, to which I referred in my last paper, and which have been described by Dr. Allen Sturge<sup>1</sup> under the name of spondylitis deformans ; but these are easily distinguished by the lower limbs being naturally straight and the clavicles and skull unchanged.

<sup>1</sup> 'Trans. of the Clinical Society,' xii, p. 204, 1879.