number of cases of dysbasia angiosclerotica, and showed that of 38 men who suffered from that disease, 10 were heavy smokers, and 15 were enormously heavy smokers; so that 25 smoked to excess out of a total of 38. More convincing still is his statement that in 14 of these cases excessive smoking was the sole etiological factor; all other possible causal agencies, such as syphilis, alcohol, diabetes, etc., were definitely excluded. Klemperer²⁵ records two well-marked cases of arterio-sclerosis which he attributes solely to excessive smoking. Neither of these men had suffered from infectious disease, nor from syphilis. They were both abstemious, and they had not been subjected to any special mental strain. One of them, aged 37, had never smoked less than thirty cigarettes, and often as many as one hundred, daily for twenty years. The left ventricle was hypertrophied; the second sound was strongly accentuated, and there was a murmur over the aorta. The radial arteries were hard and tortuous, and the pulse was of high tension. The other man, aged 44, had smoked from six to twelve cigars daily from the age of 19; between his 30th and 40th years he had never smoked less than ten cigars daily. He had suffered from a feeling of oppression of the heart for three years, and during the last six months had been attacked by severe angina pectoris. The left ventricle was moderately dilated and the second aortic sound was greatly accentuated. The radial arteries were hard and tortuous.

Klemperer states that he has seen a number of cases of arterio-sclerosis in heavy smokers, but that in these cases other factors—especially severe mental strain—were concurrently present. Muskat²⁶ enumerates the various causal factors of angiosclerosis as a forerunner of spontaneous gangrene, and says: "The abuse of tobacco stands prominently to the front." Simon²⁷ records a case of dysbasia angiosclerotica in a man aged 64 who had not had any severe nerve strain; had not been unduly exposed to the weather; had led a quiet life; had not had syphilis, nor any serious illness, except inflammation of the caecum eight years previously, but who had smoked thirty strong cigarettes daily since the age of 19; he had inhaled and swallowed the smoke. The patient complained of heaviness and weakness in the legs, headache, and cardiac disturbances. The radial arteries were hard, tortuous, and sclerosed, and the pulse was of high tension. The heart was not dilated; the second aortic sound was accentuated. Pulsation in both arteries of the left foot was entirely absent; and was absent in the posterior tibial, and only feebly present in the dorsalis pedis of the right foot.

Prolonged, excessive smoking may lead to sudden painless attacks of cardiac failure, which in some instances are fatal. One such case I saw was that of an active man of about 45 years of age, who had long suffered from indigestion and flatulence. A few days previously he suddenly had an attack of syncope without obvious cause, and fell in the street; on recovery of consciousness there was a tendency to relapses when he attempted to rise from the horizontal posture. When I saw him he was quite conscious, and was free from pain and discomfort, except that which was caused by the dread of a return of the syncope and of immediate death. The pulse was rapid and of extremely low tension; the heart sounds were well-nigh inaudible. The heart was much dilated, and no impulse could be felt nor any murmur heard. The patient told me that he had not suffered from any illnesses beyond the indigestion, and that he had been extremely abstemious as regards alcohol all his life. On being questioned, he frankly admitted that for many years it had been his habit to smoke daily "any number of cigars from ten upwards." His business involved frequent railway journeys, and when he left home very early in the morning he often smoked a cigar in place of having breakfast. The radial arteries were hard, and the vision showed the usual defect for red and green. The patient died shortly after from heart failure.

Against the view that excessive smoking can produce the results above described, it has been urged that very many heavy smokers live to advanced old age. This is undoubtedly true; but, apart from the greater evils, how many of these excessive smokers escape dyspeptic troubles, flatulence, capricious appetite, irritation of the throat, attacks of insomnia with exaggerated cardiac action, and unaccountable periods of gloom and depression? As regards longevity and immunity

from vascular changes, individuality and hereditary transmission play an important part. When discussing the objection that many heavy smokers are often long-lived, Klemperer observes that the same objection may be raised against the alleged injurious effects of alcohol. Do not many habitual drunkards live to an advanced age? Do we not frequently see alcoholics die from cirrhosis of the liver without any calcification of the arteries? Some families display a tendency to arterial sclerosis; if such a tendency exists, it is undoubtedly developed by the abuse of tobacco.

It is obvious that with all physiologically active substances the question of degree is largely determinative of the results produced by any agency that is capable of affecting the health. Most men, if they choose to smoke, can do so within certain limits without injuring their health; some men can exceed such limits with apparent impunity. The extent of the limitation must be determined by each man for himself, and if he is wise he will keep well within the border-line and will hold to his resolution. Here lies the difficulty; the growth of a habit overrides discretion. The habit of excessive smoking is more insidious in its development than that of excessive drinking, for the results are much less obvious. An alcoholic may be appealed to by his friends or admonished by those in authority; this rarely happens to the adult smoker, unless he consults his doctor about "indigestion and palpitation" and admits that he smokes to excess; then probably the necessary warning will be given.

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NOTES OF A CASE OF OSTEITIS DEFORMANS.

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[WITH SPECIAL PLATE.]

THE patient whose case is here described and illustrated by photographs and skiagrams was exhibited in the Surgical Section of the Annual Meeting of the British Medical Association, 1908. The man, who is 59 years old, a glass-cutter by trade, and of abstemious habits, came under my observation at the Sheffield Royal Infirmary a few months ago. His large cranium, kyphotic spine, and curved limbs presented a striking picture which arrested immediate attention.

There is nothing noteworthy in his family history, and he was quite healthy until the age of 20 years, when he suffered from a running ear and had an abscess at the right side of his lower jaw, the scar of which is still visible. A year later he had an ulcerated throat which lasted for two months. He denies the possibility of his having had venereal disease.

For the succeeding twenty-five years he enjoyed good health and worked regularly at his trade, but he noticed that his head was very gradually increasing in size. At 25 he wore a 6½ hat, at 35 the size had increased to 7½, and after the age of 40 years he had to have one made specially for him.

At the age of 46 a large ulcer appeared on the front of his left leg. His medical attendant, Dr. C. S. Kilham, of Sheffield, noticed at this time the deformities of his head and limbs, and told him he was suffering from Paget's disease. This was confirmed by my colleague, Mr. Charles Atkin, under whose care he became an inmate of the Sheffield Royal Infirmary. After a course of potassium iodide the ulcer healed, and he returned to work and remained in regular employment until the spring of the present year.

He thinks his limbs have become more curved and heavier during the last few years, but beyond feeling less

inclined than formerly for long walks he is not aware of any failure in his health or strength. He eats and sleeps well; has not at any time had pains in his limbs or head; his eyesight is good; and all his viscera appear to be healthy.

The accompanying photographs and skiagrams render a detailed description of the bone deformities almost superfluous.

The head is dorsi-flexed on the spine, and the chin protrudes unduly. The cranial vault is uniformly enlarged and in its greatest transverse circumference measures 24½ in. The outer surfaces of the cranial bones are studded with small pits, which can be easily felt through the scalp. There is a small exostosis on the right malar bone, but the other facial bones are unaffected. The spine presents a well-marked kyphotic curve, which involves the dorsal and lower cervical vertebrae. The movements of the individual vertebrae on one another are somewhat restricted, but there is no rigidity. The clavicles are slightly thickened and their normal curves exaggerated.

The chest is compressed laterally, so that the antero-posterior diameter is greatly increased, especially below. The ribs are unduly oblique, and the tips of the three lowest are on a level with the iliac crests. The abdomen presents a deep transverse furrow midway between the umbilicus and pubes. The pelvis in appearance resembles that of the female, the iliac bones being wide apart. The femora are much thickened and curved, with the convexity forwards and outwards. The angle formed by the neck and shaft is reduced to about 100 degrees, and the hip-joints cannot be fully extended. The tibiae are enormously thickened, especially in the lower half, and markedly curved, with the convexity in front and slightly to the outer side. The sharp anterior borders are obliterated, and the shafts are more or less cylindrical in outline. The outer surfaces of the tibiae, and to a lesser degree those of the femora, are covered with minute nodules and depressions. The tibiae are increased in length as well as in thickness. The fibulae are thickened and slightly curved but do not appear to be appreciably lengthened. This inequality in the longitudinal growth of the two bones is responsible for a portion of the tibial curve. The knee-joints cannot be fully extended. When the patient lies on his back the entire weight of the legs is borne by the heels and buttocks, and there is an interval of quite 6 in. between the backs of the knees and the couch. When the ankles are in apposition the knees are 10 in. apart. A deeply-pigmented scar on the lower part of the left leg indicates the site of a former ulcer. The bones of the upper arms are not appreciably altered but those of the forearms are enlarged and curved, with the convexity towards the dorsum. The bones of the hands and feet do not appear to be affected. According to the patient's statement, the bones of the left side in every instance became enlarged earlier than those of the right.

The late Sir James Paget was the first to recognize and describe osteitis deformans as a definite disease. His description of it, to which is appended Mr. Butlin's account of the pathological changes in the affected bones, was published in the *Transactions of the Royal Medical and Chirurgical Society of London*, 1877, vol. xlii, pp. 37-64. After relating particulars of five cases, he goes on to state that:

It begins in middle age or later, is very slow in progress, may continue for many years without influence on the general health, and may give no other trouble than those which are due to changes of shape, size, and direction of the diseased bones. Even when the skull is hugely thickened and all its bones exceedingly altered in structure the mind remains unaffected.

The disease affects most frequently the long bones of the lower extremities and the skull, and is usually symmetrical. The bones enlarge and soften, and those bearing weight yield and become unnaturally curved and misshapen. The spine, whether by yielding to the weight of the overgrown skull, or by changes in its own structures, may sink and seem to shorten with greatly increased dorsal and lumbar curves; the pelvis may become wide; the necks of the femora may become nearly horizontal, but the limbs, however misshapen, remain strong and fit to support the trunk. In its earlier periods, and sometimes throughout all its course, the disease is attended with pains in the affected bones, pains widely various in severity, and variously described as rheumatic, gouty, or neuralgic, not especially nocturnal or periodic. It is not attended with fever. No characteristic conditions of urine or faeces have been found in it. It is not associated with syphilis or any other known constitutional disease, unless it be cancer.

Paget describes at length the *post-mortem* appearances found in one of his cases. All the cranial bones were greatly thickened and the sutures obliterated. The frontal

bone varied in thickness from 11 to 13 lines, the parietal from 14 to 16 lines, and the occipital from 8 to 12 lines. The whole outer surface of the skull-cap was finely porous and perforated with innumerable apertures for blood-vessels. The outer table was represented by a mere shell of compact bone, which enclosed cancellous tissue in various degrees of fineness, the interstices being occupied by a soft reddish substance. The shafts of the long bones were in every part increased in thickness, and presented on section a finely reticulate appearance, like very fine coral. Their outer surfaces were irregular and finely nodular, and visibly perforated for the transmission of blood vessels, the appearance indicating a greatly increased vascularity of the bones.

As is indicated by the nomenclature which he adopted, Paget believed the disease to be a chronic inflammatory affection of the bones, and almost all subsequent writers have endorsed this view.

The pathological changes may be briefly summarized as:

- (a) A rarefying osteitis, particularly of the compact bone; and
- (b) The formation of new bone from the periosteum.

Lancereaux suggests that the new bone formation is an attempt at reinforcement of the long bones which have been weakened by absorption. Its distribution around the entire circumference, however, differs from other instances of compensatory growth—notably in rickets—where the reinforcing bone is developed on the concave side of the shaft. The newly-formed bone is imperfectly ossified and softer than normal bone. The increased thickness of the cranial bones is due entirely to periosteal growth, and as the cranial space is not encroached on, there is absence of cerebral pressure symptoms.

Very little is known of the etiology of the disease. In the vast majority of cases it begins during or after middle life. In a series of cases tabulated by Drs. Packard and Steele the average age at the time of onset was 49½ years, and the average duration of the symptoms at the time when the patient first came under observation eleven years. Moizard and Bourges report a case in which the disease manifested itself at the age of 21 years, and in the case I am recording the patient's head had begun to enlarge before the age of 35 years.

Three of the five cases recorded by Paget developed cancer late in life, and he not unnaturally thought there might be a relationship between the two diseases. An analysis of the published cases, however, does not support this view. Indeed, apart from Paget's group of cases, the incidence of cancer is less than one would expect, having regard to the advanced age which the majority of the subjects of the disease attain. All writers agree that it has nothing to do with syphilis. In the case I am recording, however, the evidence of syphilis is fairly conclusive.

The disease must be very rare. Owing to the remarkable deformities to which it gives rise, and to the fact that patients suffering from it usually live many years after its onset, few cases are likely to go unrecorded, and yet during the thirty years that have elapsed since the appearance of Paget's paper not more than 100 cases have been reported. A very distinguished professor of surgery, attached to one of the largest hospitals in the kingdom, with an experience of forty years, informed me that my case was the only living example of the disease he had seen.

Packard and Steele¹ have tabulated statistics bearing on the incidence of the disease as it affects the sexes and the various bones in 67 cases. Of these, 41 were males and 24 females, while in 2 the sex was not stated. The individual bones were affected as follows: The skull in 49 cases, the tibiae in 47, the femora in 40, the spine in 31, the clavicles in 24, the pelvis in 21, the ribs in 16, the humeri in 14, the radii in 11, the fibulae and ulnae each in 10, the patellae in 8, the sternum in 7, the scapulae in 6, and the metatarsi in 5 cases.

The progress of the disease is always slow, and in the majority of cases reported it does not appear to have appreciably shortened the span of life.

There is no evidence that it has been influenced by any plan of treatment hitherto adopted.

I am indebted to Dr. Rupert Hallam, Medical Officer in Charge of the Electrical Department at the Royal Infirmary, for the skiagram.

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