DISEASE OF MANY JOINTS, PROBABLY OF SYPHILITIC ORIGIN.

BY

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G. M—, æt. 16, first came under the care of Mr. Morrant Baker in St. Bartholomew's Hospital on November 20th, 1882, on account of ulceration of the left leg.

His history was that he had been in good health until three years before admission, and that about that time he had an attack of synovitis of the left knee for which he was under treatment in the London Hospital for two or three weeks. About a year later swellings began to form on his shin bones, and after several months one of these swellings burst, and ultimately some bone came away. The open sore thus formed had not healed.

On examining the patient there was found an irregular ulcer extending along the shaft of the tibia for a distance of about two inches. The edges were heaped up, and the

base of the ulcer was formed by rough, bare bone. The tibia itself was thickened.

On the middle of the right tibia and a little to its inner side was a hard nodular swelling which was neither painful nor tender. There was no other evidence of congenital syphilis, the eyes and teeth being normal.

The patient was treated with iodide of potassium and mercury, and on December 16th some of the rough carious bone was chiselled off the left tibia. This operation was repeated on February 28th, and by March 30th the leg was healed. At the time of operation the bone was found to be hard and dense.

Rather more than a year later, on June 26th, 1884, the patient returned to the hospital. The scar on the left shin had broken down, and the right shin also was now ulcerated, the right tibia being enlarged, and rough carious bone being exposed in the floor of the ulcer.

The treatment adopted was the same as that which had been found efficacious formerly, but the ulcers proved most intractable, and it was only after six months' rest in bed and several gouging operations that the shins again healed.

The patient was then sent to the convalescent home, and whilst there, for the first time since he had been under notice, the joints began to show signs of inflammation, and a sharp attack of synovitis of the left knee and of both elbows necessitated his return to the hospital. The affected joints contained a good deal of fluid and were a little painful, but under treatment by Scott's dressing the fluid was quickly absorbed, and the patient was again discharged in March, 1885.

On September 1st of the same year he was again admitted. He said that he had noticed swelling on the left side of his forehead and on his right arm for about three weeks, and that his shins had broken out again soon after he was last discharged.

Examination showed a swelling attached to the right humerus about its centre, on the outer side of the arm.

The skin over it was red and hot, and the swelling itself fluctuated. The bone around felt thickened. Over the left eyebrow was a swelling as large as a walnut, firm, and attached to the subjacent bone. In addition to the ulceration of the shins and the thickening of the tibiae there was a good deal of effusion in each knee, and some stiffness in the movement of these joints. There was also effusion in the left elbow. The affected joints gave no pain.

On September 4th pus and caseous matter were evacuated from the swelling on the right humerus and from that over the eyebrow. In each case bare and rough bone was exposed.

The general health of the patient was very bad. He was feeble and emaciated, and had continuous slight pyrexia. The sores on the shins continued to discharge, although from time to time they healed in places.

In December the patient was troubled with cough, and effusion occurred in each pleura. The liver and spleen became much enlarged and caused a general swelling of the whole abdomen. The enlargement was evidently due to amyloid disease.

The incisions over the humerus and frontal bone did not heal, and at the end of January, 1886, a swelling appeared over the left acromion and another below the body of the lower jaw on the left side.

On February 15th albumen was found in the urine for the first time, and on February 22nd the patient died.

A post-mortem examination showed the following conditions :

Head.—A small ulcerated aperture was seen over the inner angle of the left orbit, leading to bare bone. On reflecting the scalp some cheesy, gummatous-looking material was found around the bare bone. Over the posterior part of the right parietal bone, near its inferior angle, was a soft mass of gummatous-like structure about the size of half a walnut, and in connection, below, with rough and bare bone. A similar but smaller and

firmer mass was found over the left parietal bone: part of it was removed for microscopical examination. On removing the calvaria the membranes were found adherent opposite the largest gummatous mass, and the inner surface of the left frontal bone, beneath the ulcerated aperture already mentioned, was in a carious state over an area of about half a square inch. The outer table at the same spot was roughened, pitted, and ulcerated. The brain appeared natural.

Lower jaw.—Some caries of outer table opposite incisors, and of inner table opposite second molar.

Throat.—A considerable amount of cicatricial tissue was discovered in the posterior wall of the pharynx, close to the upper opening of the œsophagus.

Thorax.—Heart and pericardium, lungs and pleuræ quite natural.

Abdomen.—Greatly distended. The peritoneal cavity contained about four or five pints of clear ascitic fluid. The greater part of the abdomen was filled by an enormously enlarged liver and spleen.

The *liver* weighed 10 lbs. 9 oz.; its surface was scarred, especially on the left side and behind, and was puckered and contracted as though by condensation of fibrous tissue; its edges were rounded. Its section was firm and of the colour of flint, being mottled. The addition of iodine gave a marked amyloid reaction. The liver contained no gummata, nor did its section exhibit traces of healed gummata in any part. The cicatricial tissue did not penetrate deeply into the gland.

The *spleen* was enormously enlarged. Its shape was peculiar, for the notch was quite obliterated, and the whole gland was an evenly-shaped oval mass. The surface was neither puckered nor scarred. The section was firmer than natural, yet not very firm; the colour was unusually dark, and there was no indication of amyloid material to the naked eye, and no "sago-grain"-like appearance. The addition of iodine did not give any amyloid reaction.

The *kidneys* were large and pale, but did not give the amyloid reaction to iodine.

The *intestines* were slightly amyloid in patches.

Stomach, bladder, and pancreas normal.

On microscopical examination much amyloid degeneration was found in the liver and spleen, and very slight amyloid change in the Malpighian capsules of the kidneys.

Right upper extremity.—On the outer side of the right arm was an irregular ulcerated aperture in the skin exposing bare and rough bone. The right humerus after removal showed the following conditions. The articular cartilage of the head was thinned in almost its whole extent, and of a bluish colour. On the posterior aspect of the head, near the anatomical neck, was a deeply-cut groove, extending for an inch or more towards the centre, after which it turned towards the great tuberosity, and ramified over the greater part of the posterior surface of the head. The posterior portion of the articular surface looked as if portions of cartilage had been gouged away, so as to leave irregular tracts with crescent-like margins. Islands of cartilage here and there remained intact. At those parts where the cartilage was most deeply destroyed the ulceration had involved the bone, and it had undergone the same gouging process as the cartilage. Crossing the bone, and closely attached to it, was a thin membranous layer. When this was peeled off, the bone was found to be rough and softer than usual. Microscopically this membrane was cellular, and was continuous with the synovial membrane, fading gradually as it approached the centre of the cartilage. In other parts the cartilage was thickened and lumpy, its thickness being at least twice as great as that of normal cartilage. The synovial membrane was more vascular, and was thicker than usual, but otherwise appeared to be normal. The shaft was thickened and misshapen by the deposit of porous new bone. The new bone was in parts tolerably firm, but in other parts was soft and crumbling. Beneath the periosteum in some parts were deposits of inspissated

pus. The lower two-thirds of the shaft were more affected than the upper third, and at the centre of the shaft, at a place corresponding to the external ulceration, there was considerable destruction of bone with the formation of a deeply-excavated cavity.

Right elbow-joint.—There was a considerable quantity of thick synovial fluid. Part of the cartilage of the trochlear surface of the humerus had been converted into fibrous tissue, but on the anterior part of the same surface there was a nodular proliferation such as is met with in osteo-arthritis. The cartilage of the capitellum was fibrillated and pitted, but the bone was not exposed.

The posterior surface of the right olecranon fossa was rough and carious, and the periosteum was separated by broken-down inflammatory products of a caseous nature. On the articular surface of the olecranon the cartilage was in part destroyed, and the bone exposed and roughened. The articular surface of the coronoid process was in a similar condition, and between the olecranon and the coronoid processes the bone was grooved longitudinally in the manner commonly seen in osteo-arthritic joints. The shafts of the radius and ulna were healthy.

Left upper extremity.—The left shoulder-joint was next removed. It contained some inspissated synovial fluid. The cartilage of the glenoid cavity was normal, and the scapula did not seem to be diseased.

The shaft of the humerus was normal, but the head presented appearances similar to those seen in the other shoulder. An examination showed that the articular cartilage was thinned over a large extent of its surface, the thinning being less marked at the centre than towards the circumference. The thinner portions were of a bluish colour. At the lateral and posterior margins of the head, near the anatomical neck, were deeply excavated tracts. The lateral excavation commenced at the margin of the head and extended forwards towards the centre of the articular surface. The base of the excavation was covered by a fibrillated membrane, which was continuous with the

thickened synovial membrane of the joint. A process of the synovial membrane extended forwards and accurately fitted into the excavated surface, though it was not adherent to it. The excavation on the posterior surface of the head was more irregular, and had exposed roughened bone. From the lower portion of this surface the excavation extended downwards for a short distance along the shaft into the substance of the bone itself. The synovial membrane was thickened and fringed as in cases of osteo-arthritis.

The left elbow-joint and the left radius and ulna were normal.

Lower extremities.—The shins were covered with scars which were adherent to the bone beneath. The tibiæ were thickened and covered with porous new bone. From the front of each tibia a considerable quantity of bone had been removed. The ankle-joints and the joints of the feet were normal.

The right knee-joint contained a considerable quantity of viscid fluid, and on being opened showed the following conditions. The synovial membrane was everywhere thickened and hypervascular. The patella was surrounded by a mass of fringes, some very pedunculated, others almost sessile; some large, and resembling masses of fat, others delicate and filamentous. The external condyle presented a deep groove, which runs antero-posteriorly for about an inch and a quarter. The groove extended down to and involved, but did not expose, the bone, which was covered by a membrane similar to that which lined the "ulcerated" or grooved portions in the shoulder-joint. Towards the upper and anterior part the groove became shallower, and an island of cartilage remained intact. The cartilage on the upper part of the condyle was rough and fibrillated, and greatly increased in thickness. On the upper part of the internal condyle was a large nodular outgrowth of cartilage about an inch in length. On the most convex part of the same condyle was a small mass of fibrillated cartilage. With these exceptions the rest of

the cartilage on this condyle was normal. The cartilage of the patella was rough and fibrillated, whilst that covering the upper extremity of the tibia was slightly roughened and fibrillated on the surface. The ligaments were normal.

Left knee-joint.—The cartilage of the external condyle near to the intercondyloid notch was deeply gouged in the manner already described, as seen in the opposite knee. The destructive process had extended in a somewhat serpiginous manner, and had left islands of cartilage intact, but almost the whole of the cartilage of the anterior surface of both the external and the internal condyles had been removed, and much of the subjacent bone excavated. That part of the articular cartilage which had, during life, been in contact with the tibial tuberosities was for the most part natural, but in places was a little fibrous and thickened by the growth of small nodular masses. The bottoms of the grooves were everywhere covered by a membrane, as was the case in the opposite knee. At the edges of the condyle were nodular outgrowths of cartilage, such as are commonly seen in osteoarthritis; none of them were of large size: the external condyle was “lipped” in a very marked manner. The cartilaginous surface of the patella was rough and fibrous, but there were no outgrowths round the edge. The cartilage of the tibia was slightly roughened, and at one spot over the internal condyle, had been replaced by a soft mass of pulpy tissue, which intervened between the free surface and the subjacent softened bone. The cartilage covering the outer articular surface of the tibia was normal except beneath the external semilunar cartilage, and here its surface was fringed and rough, and in parts slightly worn away. At its edge there was a little nodular thickening, but no definite lipping.

A microscopical examination of the affected joints showed the following conditions. A section through the articular bone and its cartilage exhibited a very extensive enlargement of the cancellous spaces, with a corresponding destruction of the bony trabeculæ. The place of the bone

which had been destroyed was occupied in part by masses of fibrous tissue rich in cells, and in part by a more purely cellular growth. There had evidently been a rarefying osteitis with formation of fibrous tissue in the cancellous spaces.

Immediately beneath the cartilage, where the bone had been destroyed, there was an extension of the cell-growth into the hyaline matrix, with destruction of the latter to a varying extent in different parts. Here and there were seen small excavations in the cartilage, whilst in other parts the whole substance of the cartilage had been destroyed.

The place of the cartilage thus destroyed was taken by fibrous tissue, which was continuous with that found in the enlarged cancellous spaces of the bone ; and where the deep pits in the cartilage extended the whole of its depth and involved the bone, the thin membrane alluded to had evidently been formed by the same inflammatory process which had been in progress in the subjacent bone.

The case here recorded is, taken as a whole, unlike other cases recorded in English literature, and I am not acquainted with a similar one published abroad. Indeed, the peculiar characters of the morbid changes, the multiplicity and extent of the joint lesions in the case under consideration, and the concurrence of extensive bone lesions seem to place it in a class by itself.

With regard to the nature of the affection, I am disposed to regard it as syphilitic, and due to an inherited disease. The points in favour of this conclusion are (1st) the appearance of the affected bones, and especially the gummatous lesions in the calvaria, and the caries and thickening of the humerus ; (2nd) the occurrence of extensive scarring in the pharynx, evidently due to previous ulceration ; (3rd) the scarring and thickening of the capsule of the liver. I am inclined to consider that these positive evidences outweigh the absence of some of the more common lesions of congenital syphilis, but it should be mentioned that Mr. Morrant Baker, under whose

care the patient was during almost his whole illness, was not convinced that the conditions were due to syphilis, and was rather of the opinion that the latter was not the cause of the disease. Mr. Baker laid particular stress on the failure of the usual remedies to promote at any rate some improvement, on the marked absence of any syphilitic affections of the eyes or teeth, and on the complete want of any evidence of syphilis in the brothers and sisters of the patient or in his parents. In favour of the syphilitic origin of the trouble it may perhaps be urged that the amyloid changes in the viscera were also of syphilitic origin. I think this is very possible, but in the presence of ulceration of the shins of long standing it may well be maintained that the amyloid disease was due to chronic suppuration.

But even if the disease be considered as due to syphilis it must be confessed that the character of the joint affection is most unusual.

In vol. xxvi of the 'St. Bartholomew's Hospital Reports' I have published several cases of syphilitic disease of the joints, and these fall naturally into three classes ; of (a) synovitis with effusion, (b) gummata of the synovial membrane and perisynovial gummata, (c) syphilitic periosteitis of the articular ends of the long bones. Such cases are well recognised by writers on syphilis, as are also the still more common cases of painless effusion in the joints of children who have other evidences of congenital disease. But although this latter class of case is not uncommon in actual practice, I am not aware of any post-mortem examinations which show the actual nature of this affection ; and it is possible that in such cases lesions such as are seen in the specimens from the case under consideration may exist, although probably to a much less extent. It is, moreover, noticeable that, in the patient whose case I record, the painless nature of the lesions, the passive effusion, and the absence of any easily perceptible thickening of the synovial membrane very closely simulated

the conditions found in this well-recognised class of chronic synovitis in children with inherited syphilis. It is also a fact that in some at least of these latter cases treatment does not produce any definite improvement; and Mr. Jonathan Hutchinson, jun., in a lecture which I shall allude to, mentions a case where the effusion lasted for many years, whilst I have recently had under my care a little girl of twelve in whom three months' treatment has caused no diminution in the size of a pair of knees greatly distended with fluid.

With regard to the joint lesions I have described, I would draw especial attention to the nature of the affection of the cartilage. It is, in my opinion, quite different from that met with in any other form of arthritis. From a microscopical point of view, the fibrillation of the matrix and the distension of the cartilage capsules would seem to indicate a relationship to the changes seen in osteo-arthritis; but the more gross lesions do not bear out the similarity. Instead of being worn down by the rubbing movements of the joints the cartilage looks as though it had been removed in segments by a gouge, and it is noticeable that this destruction does not in any way correspond to the sites of the greatest pressure or attrition. The appearance of the diseased cartilage indeed suggests a gouging, cutting, or boring process, and Mr. Baker has suggested the term "terebrating ulceration" to indicate this most striking feature of the disease.

The explanation of the destructive process in the cartilage remains for consideration, and the question at once arises whether the process originated in the cartilage itself or was secondary to disease in the synovial membrane or the bone. The answer must be determined by a consideration of the conditions presented by the synovial membrane and the bone at the site of the lesions in the cartilage, and it is at once evident that there has been no direct extension of any inflammatory process from the synovial membrane, for there are no adherent processes of the latter extending into the pits in the cartilage, such

as are found in tubercular disease, no evidence of superficial erosion, such as may be seen in acute arthritis, and no gummatous infiltration.

On the other hand, there is good evidence that the bone at the site of the cartilage lesions is diseased, and the clinical history, as far as the lower extremities are concerned, certainly pointed to affection of the bones before the joints were involved.

It has already been mentioned in the description of the specimens that the pits in the cartilage correspond in many places with pits in the bone, and a closer examination of the latter shows that there has been a rarefying osteitis. It appears, therefore, probable that the cartilage has been involved in a process of so-called "subchondral caries," which has caused an inflammation of the cartilage spreading from below towards the surface.

Mr. Baker, however, has pointed out that there may be another explanation of the pits in the cartilage, and suggests that the pressure of the very large synovial fringes, some of which indeed appear to fit into the pits, may have caused a process of absorption, which would occur all the more readily when the subjacent bone was softened by inflammation, and when the joints were kept almost constantly in one position, as were the knees for a period of several months.

But whilst I consider that the deep erosions, pits, and grooves in the cartilage are not due to a primary chondritis, I must point out that, as I have already described, the surface of the cartilage is roughened in places where there is no deep destruction of it, and the microscope shows that this roughened cartilage is broken and fibrillated as regards its matrix, and that its cartilage cells are increased in number and distend their capsules. There is, further, a growth of nodules of cartilage forming lips around the articular margins, and these are in no way due to a subjacent bone disease. These changes in the cartilage should therefore be considered as primary, and not due to disease of the bone or synovial membrane.

Lastly, it may be pointed out that the synovial membrane itself shows no change which can be classified as inflammatory. Its thick shaggy fringes, its smooth surface, the absence of all adhesions, and the character of its secretion make it resemble in every particular the synovial membrane from a case of osteo-arthritis. There is a complete absence of anything like either gumma or tubercle.

In conclusion I would refer very briefly to any writings that seem to have a bearing on the subject.

All recent writers on diseases of joints and on syphilis have alluded to syphilitic arthritis, and have described several varieties, and it is not necessary to say of their writings more than this, that they do not describe the changes seen in the specimens under consideration. If, therefore, these are syphilitic joints, as I consider they are, they present unusual appearances.

Virchow, however, in 1884, in the 'Berliner klinischer Wochenschrift' for August 18th, describes a form of chondritis with fibrillation of the cartilage as due to syphilis, and says that such joints may simulate those of osteo-arthritis. It is, however, difficult to be sure that the specimens he saw were identical in nature with those under consideration. In my opinion, however, the changes seen by him were in all probability of the same nature as those here described.

In vol. xl of the 'Pathological Society's Transactions,' at page 67, Mr. Jonathan Hutchinson, jun., describes a specimen of disease affecting the knee-joint taken from a man aged 23, who had stricture and ulceration of the rectum, and interstitial nephritis, but no other evidence of syphilis, and no history of it. Mr. Hutchinson considered the joint to be syphilitic, and, in alluding to it in his lectures at the Royal College of Surgeons in 1892,¹ he considers it to be an example of the chondritis described by Virchow, and of the same nature as the specimens now under consideration. I would, however, point out that

¹ 'Brit. Med. Journ.,' 1892, vol. i, p. 728.

there is hardly even a superficial resemblance between the specimens described in this paper and the joint described by Mr. Hutchinson. It is only in the fibrillation of the cartilage that they are alike. In all other respects the specimens differ widely, for in Mr. Hutchinson's case there was during life the usual fixation and deformity commonly seen in destructive disease of the knee, the joint being flexed, rotated outwards, and displaced backwards, and all movements were greatly limited. The joint itself showed a general thickening and roughening of the synovial membrane, an absence of all pedunculated fringes, the presence of extensive adhesions, and a soft mass of gumma-like material in the substance of the synovial membrane. None of these conditions were present in the joints here described; and, on the other hand, the knee described by Mr. Hutchinson showed none of the characteristic cartilage lesions to which I have alluded.

I am, however, much indebted to Mr. Hutchinson for a reference to a paper by Gies in the '*Deutsche Zeitschrift für Chirurgie*,' 1881, p. 589, in which cases of thickening of the synovial membrane and the formation of fringes, with growth of nodular ecchondroses are described in connection with syphilis, and of which good drawings are supplied. I think that some of the cases described by Gies more nearly resemble my own than others with which I am acquainted, and they, together with the paper by Virchow, support the conclusion that such lesions as I have described may be due to syphilis.

(For report of the discussion on this paper, see '*Proceedings of the Royal Medical and Chirurgical Society*,' Third Series, vol. vi, p. 6.)

DESCRIPTION OF PLATE I.

Disease of many Joints, probably of Syphilitic Origin (ANTHONY BOWLBY).

FIG. 1.—The right humerus, showing gummatous ulceration of the shaft and destruction of part of the articular cartilage.

FIG. 2.—The right knee-joint, showing the synovial fringes around the patella and destruction of part of the articular cartilage of the femur.

FIG 3.—The lower end of the left femur, showing the deeply-gouged ulceration of the cartilage of the external condyle, and the overgrowth of cartilage on the anterior surface of the bone.

Fig 1

Fig. 2.

Fig. 3.