A case of osteitis deformans / by John R. Lunn.

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A CASE OF OSTEITIS DEFORMANS.[1]

By JOHN R. LUNN.

The name Osteitis Deformans has been given by Sir James Paget to a remarkable disease which he has described in the 'Med.-Chir. Transactions' of 1876. Mr. Bryant also described a case in the 'Guy's Hospital Reports' for 1877.

The disease begins, writes Paget, late in life; is somewhat slow in its progress, may continue for many years without affecting the general health, and gives no more trouble than that which is due to the changes of the shape, &c., of the diseased bones. He also states that the affection is not associated with syphilis or any other constitutional disease, unless it be with cancer.

William W—, æt. 70, single, by trade a hawker, was admitted into the Marylebone Infirmary, July, 1883. He had been in the workhouse eight years. He said he had always enjoyed good health and had never suffered from gout or syphilis. All his family were healthy as far as he could remember.

On admission he complained of rheumatic pains chiefly in the right hip and thigh. He looked pale and cachectic and was about five feet two inches high. His chin seemed lower than the top of his sternum, and his head hung down as if too heavy for his body. No deformity was observable in his face. The clavicles were slightly enlarged. Right humerus slightly twisted outwards and thickened. Spine twisted and curved

[1] The skeleton is now in the Museum at St. Thomas's Hospital.

laterally. Right femur was rotated outwards and felt much thickened; left femur seemed similarly deformed but to a less extent. Both tibiæ were slightly thickened and very prominent, especially the left. There was no apparent inequality in the length of the lower limbs. No visceral disease. No albumen in urine. The patient steadily lost flesh and became weaker; at first he could walk about, but he became by degrees quite confined to bed, generally lay on his right side, and in time bedsores formed over the sacrum. He was not able to get complete rest from the heavy aching pains and weariness in his limbs, which were rather severe and formed his only complaint. As he became weaker, he grew sleepless at night and drowsy by day, taking but little notice. His mind wandered, and he had occasional hallucinations of fear and delusions of suspicion. He finally died of exhaustion October 20th, 1883. Some weeks before he died he got œdema of his right lower limb, which was due to a tumour situated in the groin and right iliac region, and fixed apparently to the tissues around.

Post-mortem examination (fifty-four hours after death).—Body emaciated, great œdema of right lower limb. Rigor mortis passed off. Rib cartilages calcified, but soft and brittle, and easily cut.

Head.—Skull enormously thickened, from two to three times as thick as in health, rather soft and porous. Dura mater normal.

Brain shrunken, apparently from diminution of cranial cavity, for the convolutions were small and approximated, and the lateral ventricles were diminished in size. The hemispheres were symmetrical. No lesion of any part detected.

Heart.—A little fluid in pericardium. Heart much enlarged, walls greatly thickened, especially those of the left ventricle. Valves competent. Flaps of mitral and aortic valves calcified near their attached borders. Cavities contained dark, soft clots. Aorta much dilated and atheromatous.

Lungs of average size; œdema of both; a few ounces of serum in right pleural cavity.

Spleen 15 oz. in weight, large and soft on section, red, and very pulpy.

Kidneys of normal size and structure.

Pelvis.—A large brain-like tumour occupied the right

iliac fossa, springing apparently from the brim of pelvis, just above the acetabulum at junction of pubes and ilium. It was globular and smooth on its free surface and covered with peritoneum. It had displaced the iliacus muscle, and had caused absorption of much of the ilium, viz. the anterior third of the iliac crest and the brim of the pelvis adjacent, so that the outer part of the acetabulum was friable and nearly destroyed by the soft new growth. The growth extended down beneath Poupart's ligament, close in front of the pelvic bone, nearly to the lower border of the ramus of ischium, and caused a large prominence at the upper part of anterior surface of thigh. The femoral and inguinal glands contiguous to it were swollen and hard, and the femoral vein was adherent to it, swollen, thickened, and blocked for about five or six inches by a rather recent black thrombus. On section the tumour was found to be soft, fleshy, vascular, and mostly solid, though partially cystic, there being a large cyst in the upper part containing clear fluid, which probably accounted for the recent rapid enlargement of the tumour. There was a small superficial nodule on the right scapula.

Spine was seen from the front to be much twisted and curved in the lumbar region, and the vertebræ seemed broad and projected to the left. In the dorsal region the convexity was towards the right.

Right-hip joint.—Acetabulum partially destroyed; cartilage absorbed over diseased bone, and also eroded over head of femur in corresponding position. There were several small osteophytes on the neck of the bone. The right femur seemed much thickened and enlarged, as did the left though to a less extent.

Description of the Skeleton. By Mr. Charles Stewart, Curator of the St. Thomas's Hospital Museum.

The parts affected are generally increased in bulk and have assumed a light and porous character. The external surface of the diseased bone and the walls of the medullary cavities are in some places porous, but are mostly formed by a thick layer, having a dense white and ivory-like appearance. The

normal cancellous bone is replaced by one of a more dense and uniformly porous nature.

Skull.—The disease chiefly affects the frontal, occipital, and parietal bones; the maximum thickness being in the frontal and occipital regions, where the inner table measures 13 mm. and the external 9 mm., the diploe being faintly indicated. The grooves for blood-vessels on the inner surface are deepened. The base of the skull is nearly free from change. The temporal bones show some increased porosity in places but no thickening; the sphenoid increased porosity and slight thickening. The ethmoid, superior and inferior maxillæ, malar, nasal, and turbinal bones, and the vomer are free from disease. All the sutures with the exception of the squamo-parietal are obliterated.

Spinal column.—The whole length of the spine shows characteristic changes, but they are most marked in its lower half. Bony processes from the margins of the bodies arch over the intervertebral discs and are frequently blended together. There is also extensive ossification of the fibrous tissue connected with the spinous, articular, and transverse processes. By these means the sixth, seventh, eighth, and ninth dorsal vertebræ are united; the tenth and eleventh are ankylosed, ossification of the interspinous ligament forming a broad plate of bone, and the arches being completely fused; the body of the tenth is much wasted causing angular curvature. The twelfth dorsal and first lumbar are free; the second, third, and fourth ankylosed together, and the fifth ankylosed to the sacrum and innominate bones.

Ribs.—The ribs are only slightly affected, chiefly at their extremities; at their sternal ends many show ossification extending from them into the perichondrium.

Costal cartilages.—These show numerous patches of bone formed by ossification of the perichondrium.

Sternum.—Slightly rougher than normal, junction of manubrium and gladiolus still cartilaginous, ensiform process ossified.

Right scapula.—Head, coracoid, and acromion processes, free edge of spine, and axillary border, slightly affected.

Left scapula.—Same parts diseased but much more so, especially the spine and acromion; the venter is also somewhat affected.

Right clavicle not increased in size, but light and with bony outgrowths on outer third.

Left clavicle increased in size and with osteo-porotic features well marked.

Pelvis.—The fifth lumbar vertebra is ankylosed to sacrum and with it to the ilia. The transverse ligaments of the hip-joints are ossified, and numerous bony outgrowths spring from the margins and processes of the bones. A soft tumour rises from the interior of the right ilium close to the acetabulum, forming a chamber (70 mm. by 35 mm.) opening into that cavity, and appearing above the bone as a projecting mass. The plane of the ilia is abnormally flat, all parts show advanced conditions of the disease.

Right humerus.—Head and middle two thirds of shaft chiefly affected, lower extremity nearly normal.

Left humerus.—Head and upper two thirds of shaft affected, lower extremity normal. Right and left ulna and radius nearly normal.

Hands.—Slight bony outgrowths near extremities of meta-carpal bones and phalanges, otherwise normal.

Right femur.—The condyles alone appear normal or nearly so. The shaft is much increased in size, especially from side to side; at the middle it measures 62 mm. in diameter. Head and neck at right angles to shaft.

Left femur.—90 mm. of lower end not enlarged, cancellous tissue normal but surface somewhat too porous. The remainder in same condition as the right. A section shows the medullary cavity to be enlarged, its surface irregular and formed by bony buttresses and plates, having a dense, smooth, ivory-like surface; some small masses of finely porous bone are seen here and there. The cancellous tissue of head and neck are replaced by osteoporotic bone.

Right tibia.—Not increased in size, but light and with porous surface; the internal facet is depressed, and a few slight bony outgrowths roughen its upper extremity.

Left tibia.—Rather more than half the lower extremity of the shaft is increased in size, and measures 17 mm. in diameter; in front the surface is porous, but behind a thin layer of ivory-like bone covers its exterior.

Fibulæ nearly normal, surface only slightly roughened.

Patellæ light, otherwise normal.

Feet in same condition as hands.

Dr. Sharkey examined microscopically the occipital bone and the tumour from the ilium, and the following is his report:

The most striking points in sections of the occipital bone are (see Plates V and VI):

1. The irregularity in the anatomical structure of the bone.

2. Absorption or rarefying ostitis appears to be the predominant process, producing very large and very irregular Haversian canals, which present Howship's lacunæ as well as much larger indentations in their walls. They are lined by a thin layer of tissue which stains with logwood, as is usually the case in rarefying ostitis.

3. The bone corpuscles are distributed with great irregularity, presenting for the most part no arrangement in parallel lines.

4. The bone lacunæ are mostly small, shrunken, and supplied with but few processes.

5. There is but little appearance of the natural lamellar arrangement in the walls of the Haversian canals, and where lamellæ are seen they are ill defined, and many having different directions meet together in the walls of one Haversian canal.

6. There is also clear evidence of a formative ostitis, seen in the reduction of some Haversian canals to the smallest calibre, and in some parts the parallel lamellæ of new bone which have produced this change can be clearly made out.

7. Besides the lines in the walls of the Haversian canals, which are due to their lamellar arrangement, there are others quite irregular in appearance, and consisting of series of curves, which cross each other, or else follow more or less the direction of the lamellæ. The nature of these lines is not evident.

The portion of the tumour from the ileum which was given me to examine, presented the microscopical characters of cartilage throughout. Nowhere was any sarcomatous tissue seen.

Remarks.—The case just described is a good example of an interesting and rather uncommon disease. Its interest depends upon the remarkable and characteristic deformity, which is very noticeable during life, and the disease being "all on the outside," as well as chronic and comparatively painless, is well

adapted for clinical observation. The skeleton too makes a good museum specimen.

Bones from cases of this disease of more or less ancient date may be found scattered about in museums, but they have been observed in a vague and piecemeal way, and confounded with deformities due to other diseases or to accidents. Sir J. Paget (who described the condition in 1876) was the first to recognise it as a distinct disease, putting all its symptoms together, observing its course, and showing it to be distinct from previously known affections. Since his first series of cases was published, so many other instances have been recorded by different persons that the subject may be said to have lost some of its novelty. But still its essential nature remains as obscure and disputed as ever. I think, however, by looking at the phenomena of this case, a few points in the history of the disease may become clearer, even though no insight be gained into its fundamental pathology. The principles of mechanics will help to explain how the deformity arose.

The first fact that strikes the eye is the curvature of the long bones of the lower limbs, and the bending and crushing as it were of the spinal column. This is evidently due to the superincumbent weight of the body, and we may draw the conclusion that at some previous time the bones of support had lost their normal strength or solidity. If we look at the bones themselves to see how to account for their weakness, we find a definite change in their structure, sufficient, I think, to explain this yielding; in fact, a great part of the natural firm bone has disappeared, and in its place is a large quantity of porous and spongy bone. This new bone is firm enough, though clumsy in appearance, and would not be likely to bend. The most reasonable explanation is that the bones gave way and altered in shape while the natural bone was being removed, and that the latter was subsequently replaced by spongy bone.

The morbid process was nearly universal, but its chief stress seems to have fallen upon the bones about the main axis of the body, the cranial vault, spine, pelvis, and long bones of the lower limbs being most damaged, the face, fingers, and toes least. The absorption and disappearance of the original bone seems to have been the primary event in the course of the disease, and this must have taken place not by

any coarse process, but by very delicate interstitial changes. The latter have affected not only the compact tissue of the shafts, but also the cancellous tissue of the extremities of the long bones and the short bones.

The formation of the porous bone must be regarded as secondary, but how is its presence to be explained? Is it too much to assume a remedial effort of nature to compensate for the weakening of the bony supports? for the same thing may be seen in the curved bones of Rickets and in the formation of callus after fractures. In fact, the new bone of Osteitis Deformans bears a close resemblance to callus. Further evidence of this being the true explanation is seen in the irregular way in which it is distributed in places exposed to movement, strain, and friction, *e. g.* in and about the vertebræ and hips. The thickening of the cranial walls seems the most difficult to explain as a natural compensating process, but the high degree it reaches may be accounted for by the large area of periosteum in proportion to the size of the bones. The lengthening of certain of the long bones, if real and not merely relative, could of course be explained as a too copious throwing out of callus. Against the supposition that the formation of new bone is primary, is the fact that places escape ossification which are naturally the seats of it in advancing age, *e. g.* the rib cartilages and mid-sternal joint.

What can be the nature of the morbid process which produces such anatomical alterations? Sir J. Paget regards it as a kind of chronic osteitis, but I think this view is untenable without too elastic a use of the term inflammation; for there is no fever and no intercurrent attacks of periostitis or other acute inflammation occur during the course of the disease; besides, anatomically, the appearances are not those of inflammation.

In comparing osteitis deformans with other diseases, I would of course distinguish it from multiple exostosis; for in this the new bone is a mere addition not a replacement. Nor must the curvatures of Osteitis Deformans be mistaken for those of Rickets; for except that they are both due to giving way of weakened bones under strain, weight, or other pressure, the two conditions present no resemblances. Rickets being due to imperfect formation of bone during periods of growth only affects the growing ends and begins early in life; whereas

in osteitis deformans there is an atrophy of bone already long developed, and consequently the curves are in different places. The only diseases with an apparently real affinity to osteitis deformans are:

1. Atrophy of bones in general paralysis, a condition which is more or less common, but only partial, and in which the bones show brittleness but no consecutive hypertrophy.

2. The disease most nearly allied to osteitis deformans I would take to be osteomalacia, which is equally general and consists in a process of osseous absorption, probably identical in nature but more severe, so that the destruction being greater and more rapid, there is no time for the mechanical rectification of the bones by compensatory overgrowth. In a few mild cases, however (*vide* Billroth), some small scattered osteophytes are found. Both diseases are probably of constitutional origin, though their causes are quite unknown; both end at last by a fatal cachexia, though each is liable to be aggravated by certain complications.

Thus death in osteitis deformans may perhaps be accelerated at last by compression of the brain, from thickening and internal growth of the cranium, or the growth of new bone may be so active as to burst through all restraint, and result in the formation of well-defined tumours, as was the case in some of Sir J. Paget's cases as well as in the one here recorded.

If the view contended for in this paper be correct, then osteitis deformans consists of—

1. Constitutional disease producing atrophy and absorption of a large part of the osseous system.

2. Consequent weakening of the bones, so that they yield when exposed to strain.

3. Compensatory strengthening by the growth of what may be looked upon as a variety of callus.

4. The occasional formation of definite tumours.

5. A fatal cachexia.

I am indebted to my late colleague Dr. F. L. Benham for the able assistance he has given me in the preparation of this paper, as well as to my friends Mr. Stewart and Dr. Sharkey for the descriptions which they have supplied me with.

PLATES I—VI,

Illustrating Mr. J. R. Lunn's Case of Osteitis Deformans.

PLATE I.—Front view of skeleton.
II.—Back view of skeleton.
III.—Lateral view of skeleton.
IV.—A. Right femur.
B. Longitudinal section of left femur.
V.—Section of occipital bone under low magnifying power. (× 40 diam.)
VI.—Section of occipital bone under high magnifying power. (× 333 diam.)

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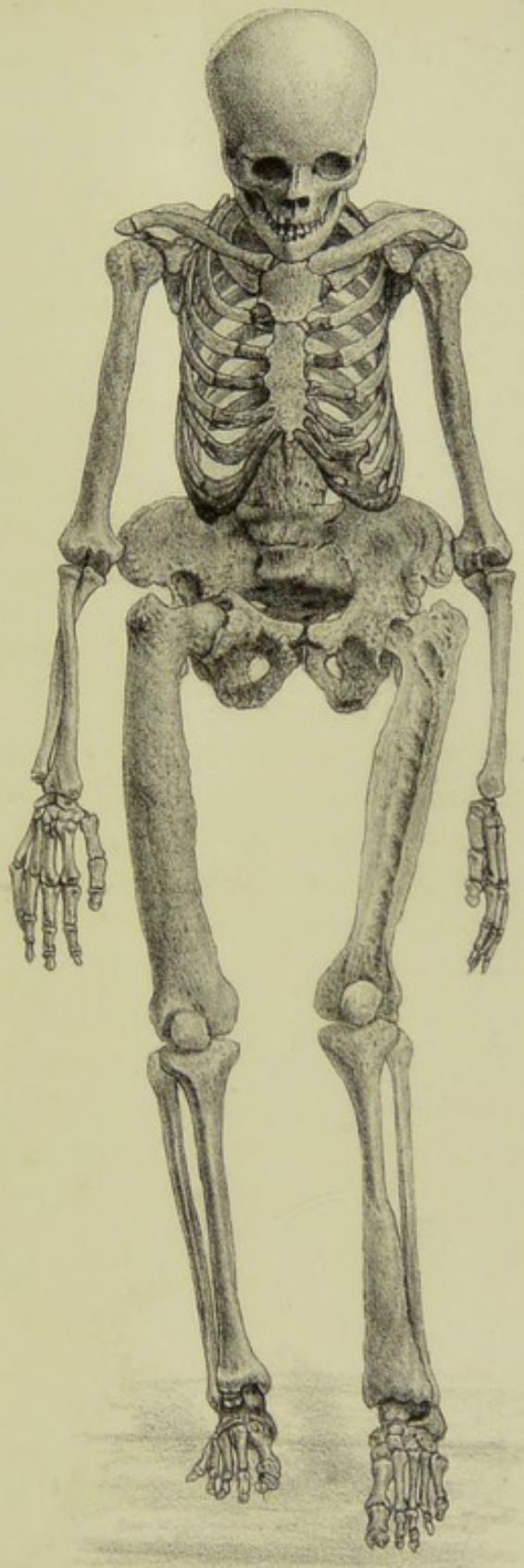

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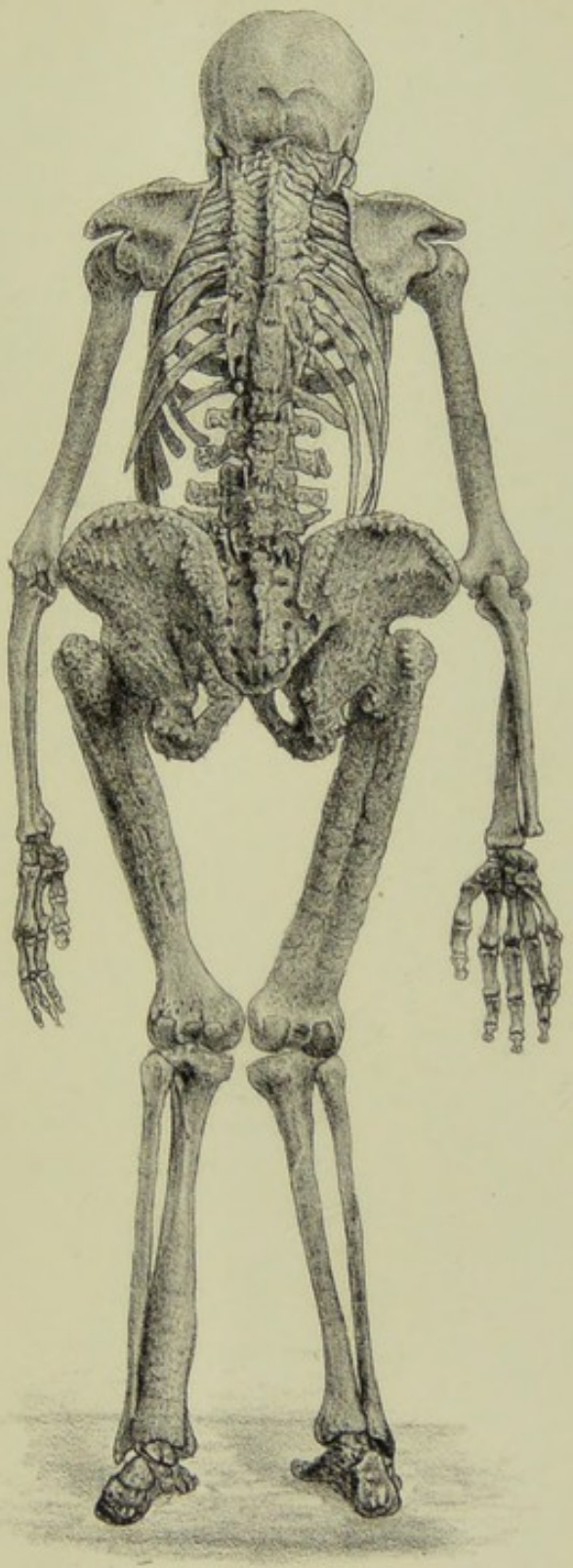

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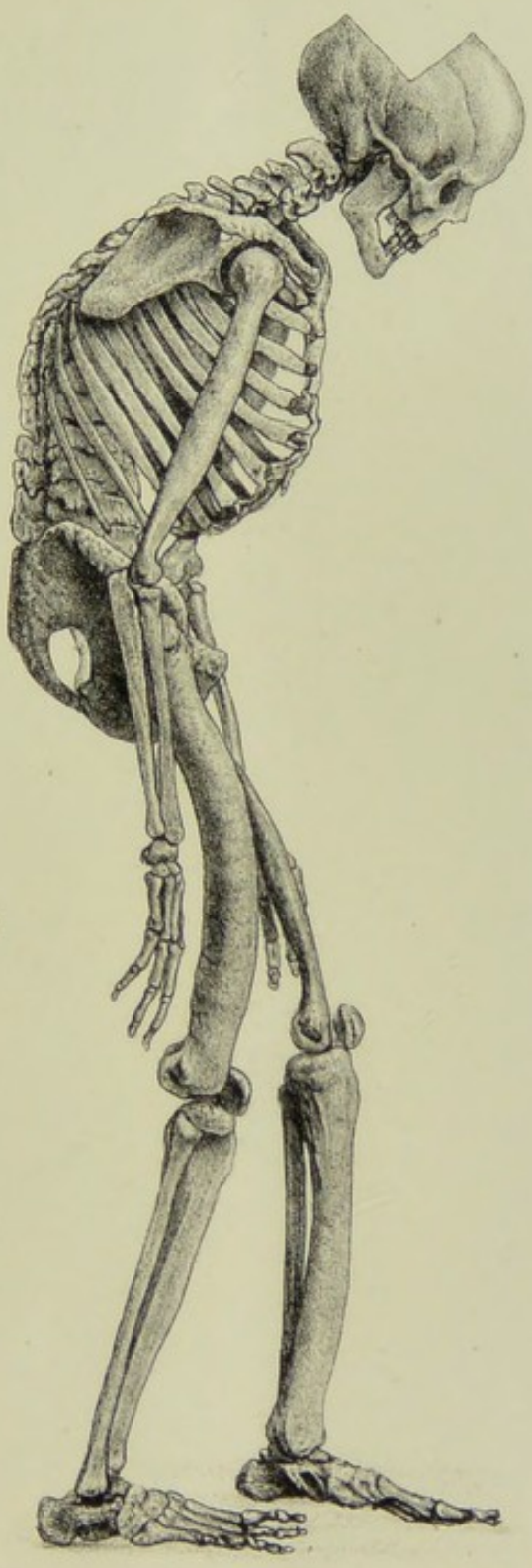

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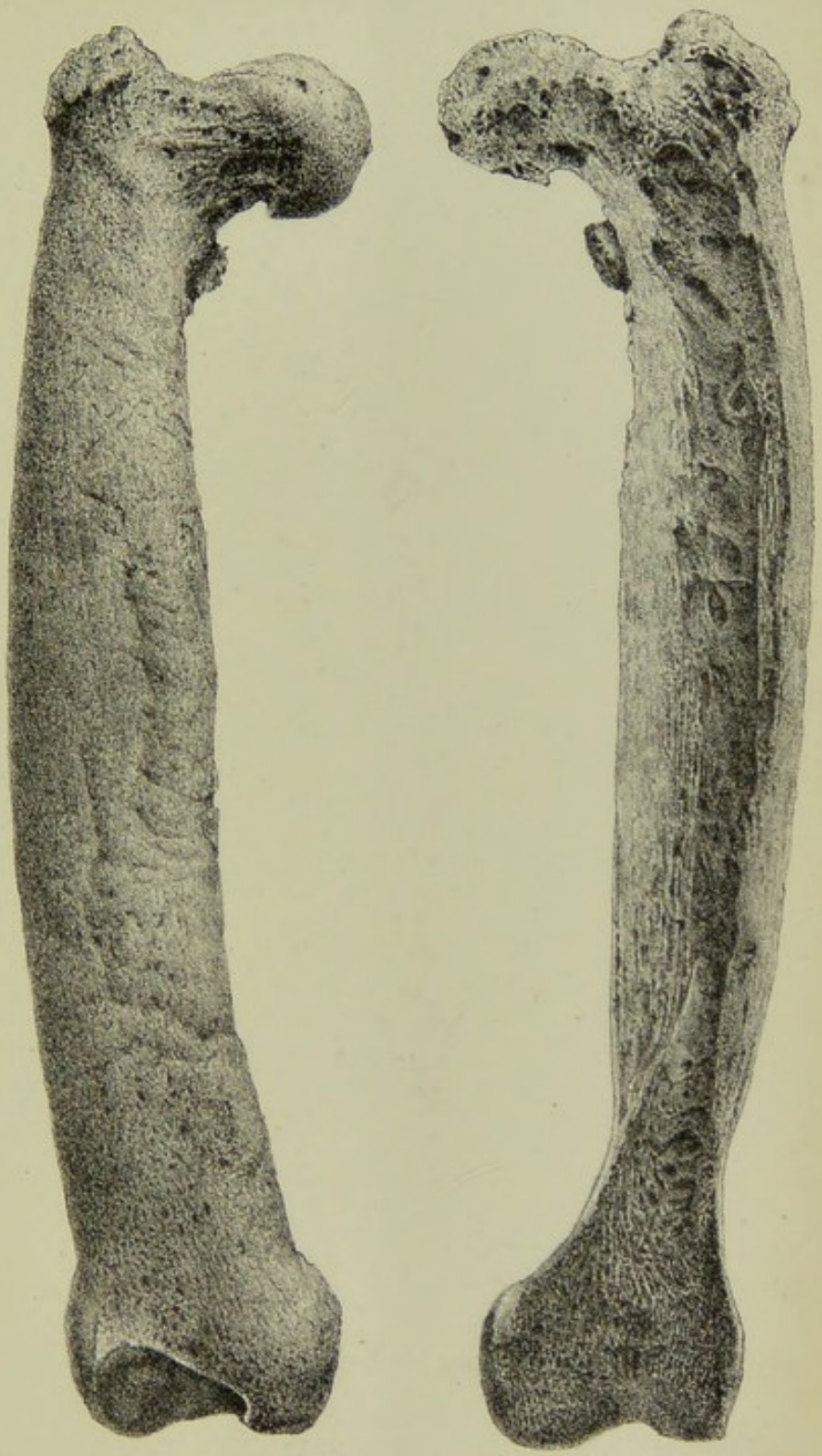

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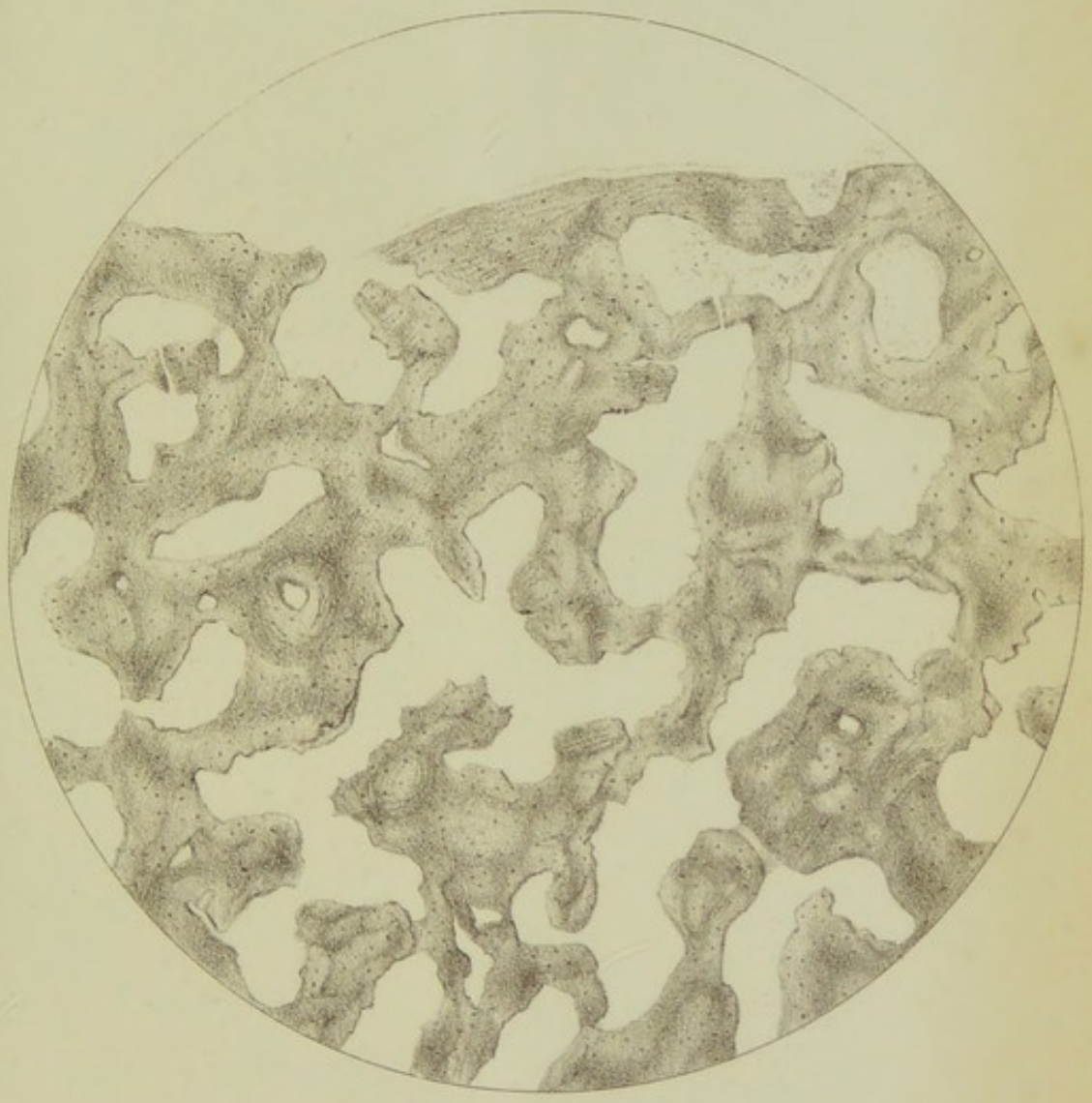

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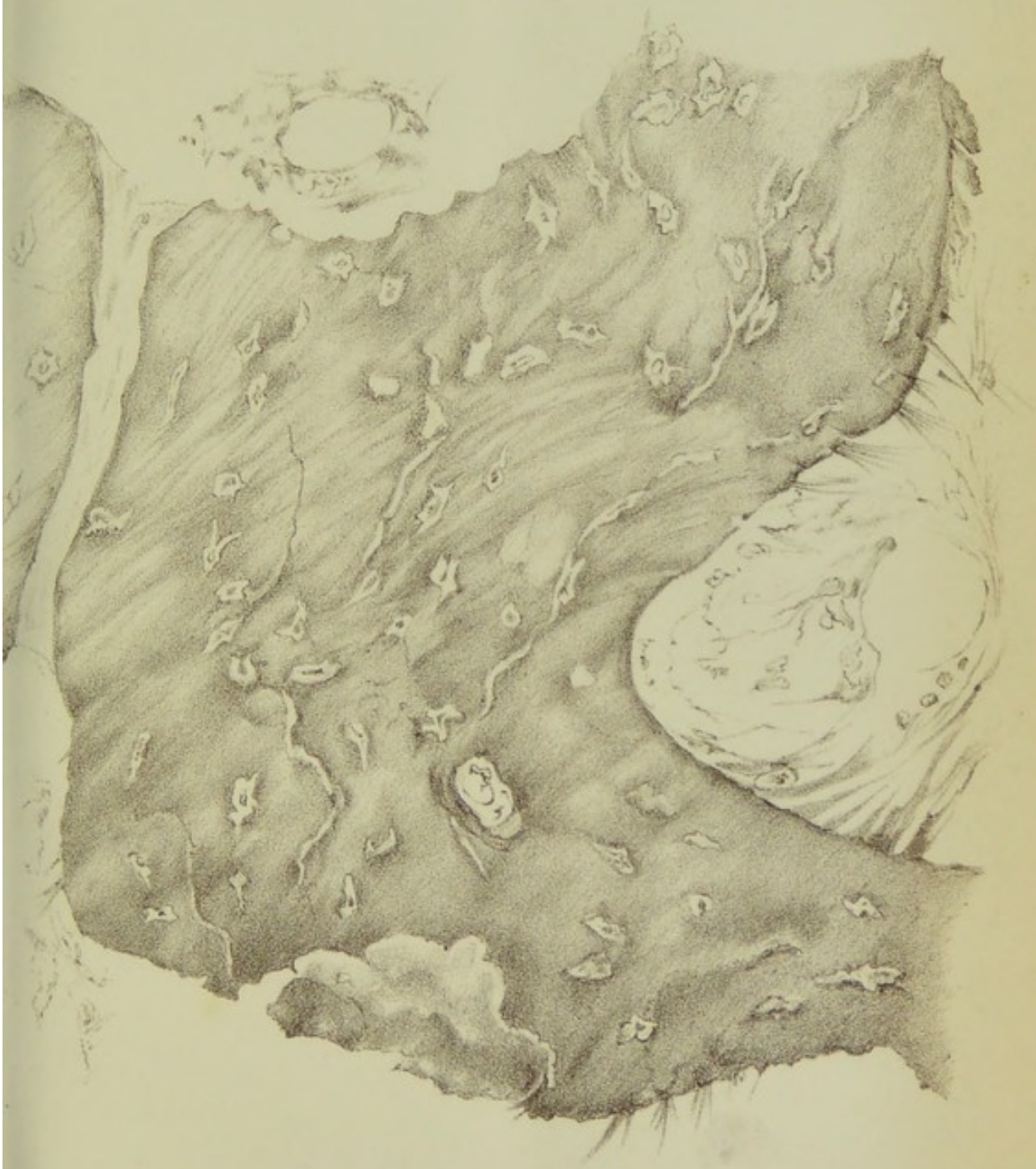

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