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NEW YORK.

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OSTEITIS DEFORMANS (PAGET'S DISEASE).*

By W. GILMAN THOMPSON, M.D.,

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Sir James Paget in his original memoir on osteitis deformans presented a study of five cases in November, 1876, before the Medico-Chirurgical Society of London, to which he added seven more in the Transactions of that society in 1882. In a review of the literature in the *American Journal of the Medical Sciences* in 1901, Packard was able to collect sixty-six cases reported to that date. This list was augmented by Higbee and Ellis who found 158 cases reported prior to January, 1911. Since that date S. F. Jones has described in the Medical Record of December 28, 1912, another case occurring in a young woman, so the case herein reported may be regarded as the one hundred and sixtieth.

This case, shown in the accompanying illustrations, is that of a woman who entered the Cornell University Medical Clinic in Bellevue Hospital in the latter part of January, 1913. She is 60 years of age; a native American; employed in general housework. Unfortunately her mind is so childish that it is impossible to obtain any history from her which is of the slightest value. For example, she

*A case reported to the Practitioners' Society, April 5, 1913.

attributes her bone deformities to fractures, claiming to have walked around with a newly fractured thigh six years ago, without surgical treatment! She admits, however, that when 15 years of age her head appeared uncommonly large, and that she ceased menstruating at 40 years of age. She appears further to have had the general pains in the extremities which characterize the early stages of the disease, but they have long ago ceased, nor has she at the present time any subjective symptoms whatever, having entered the hospital for an attack of bronchitis which completely subsided.

Upon examination the deformities of the cranium, pelvis and bones of all the limbs are very striking. The following measurements were recorded.

Total height, 145 cm.; crest of ileum to heel, 83 cm.; crest of ileum to vertex, 62 cm.; acromium to tip of middle finger, 64.5 cm.; circumference of thorax at nipple line, 74 cm.; circumference of head, 58.5 cm.; arch over vertex between ears, 31.5 cm.; arch over forehead between ears, 31.5 cm.

The patient exhibits a slight dorsal kyphosis. The pelvic brim is wide and the abdomen is protuberant and globular—the *ventre de batracien* of the French writers. On attempting to stand erect with the legs in the natural position, they present such extreme elliptical outward bowing that the knees cannot be approximated within 10 cm. and the feet are everted almost at right angles to the normal position. Hence the patient cannot balance herself in the normal erect attitude but manages to walk by crossing the right leg in front of the left, thus forming a figure-of-eight as shown in the accompanying picture, Fig. 1. The femora are markedly bowed outward and forward, after the usual manner of this disease, and the accompanying *x*-ray, Fig. 2,

shows them to be irregularly hypertrophic, being thickest in the upper two thirds, and thinnest at the junction of the middle and lower third. The humeri, particularly the left one, are in like manner bowed outward and irregularly hypertrophied, Fig. 3. The bones of both forearms and legs and the clavicles present similar enlargement and distortion, but in lesser degree. The ribs are unaffected, as well as the bones of the hands and feet. The cranial vault is exaggerated and the parietal and frontal bones especially are broad, thick, and grossly uneven on the surface. The postero-temporal and parietal regions are so prominent as to depress the ears and give the face and head a wedge shape. This is accentuated by a narrow-pointed protuberant inferior maxilla. Such an extreme case is easily distinguished from rickets. There is no deformity of the sternum, beading of the ribs, enlargement of the epiphyses or craniotabes, and the *x*-ray pictures are diagnostic in showing the irregular hypertrophic osteitis, particularly of the cranium, humeri, and femora. Furthermore, the disease has developed during adult life.

The general health of the patient is excellent. She is entirely free from pain or tenderness over the affected bones, and the detailed physical examination of the viscera reveals no abnormality excepting in the heart a slight mitral regurgitation. There is, however, advanced arteriosclerosis and the blood-pressure averages 170 mm. There is no abnormal pigmentation of the skin or hypertrophy of the soft parts such as is usually observed in acromegaly. The Wassermann reaction is negative.

None of the numerous theories to explain the origin of osteitis deformans has met with general acceptance. It is true that a majority of those who

have studied the few recorded cases, led by Lannelongue and Fournier, have favored the idea that it arises from hereditary syphilis, and the theories of its origin in trauma, a rheumatic diathesis, a trophoneurosis, or sclerosis of the nutrient arteries, have been abandoned. But the chemical analysis of the bones in syphilis and osteitis deformans is quite different, and the progress of the latter disease is not decisively modified by antisyphilitic treatment, nor is there any very definite evidence in the majority of cases that the lesions are parasyphilitic manifestations. Attempts to refer the disease to disordered functioning of the thyroid, suprarenal or pituitary bodies have similarly failed of confirmation in most of the autopsies recorded, although in one or two cases there have been found changes in the pituitary body.

Paget called attention to the coincidence of malignant disease with osteitis deformans, and five of eight patients, whose life histories he obtained, died of sarcoma or carcinoma. But this form of osteitis is very chronic and often is not established until late in life when malignant disease is common. Moreover, in a study of the 158 cases recorded prior to January, 1911, Higbee and Ellis were able to discover only fourteeen instances of malignant disease. The following year another example of the combined diseases was reported from Montreal by Gruner, Scringer, and Foster, occurring in a male 56 years of age. (*Arch. of Internal Med.*, June, 1912. Vol. 9, pp. 641-656.) In this case, however, 6 years elapsed between the appearance of the osteitis and the sarcoma and the sarcoma invaded only a few of the many bones involved in the osteitis. In this case, also, it is interesting to note, there was marked atrophy of the pituitary body.

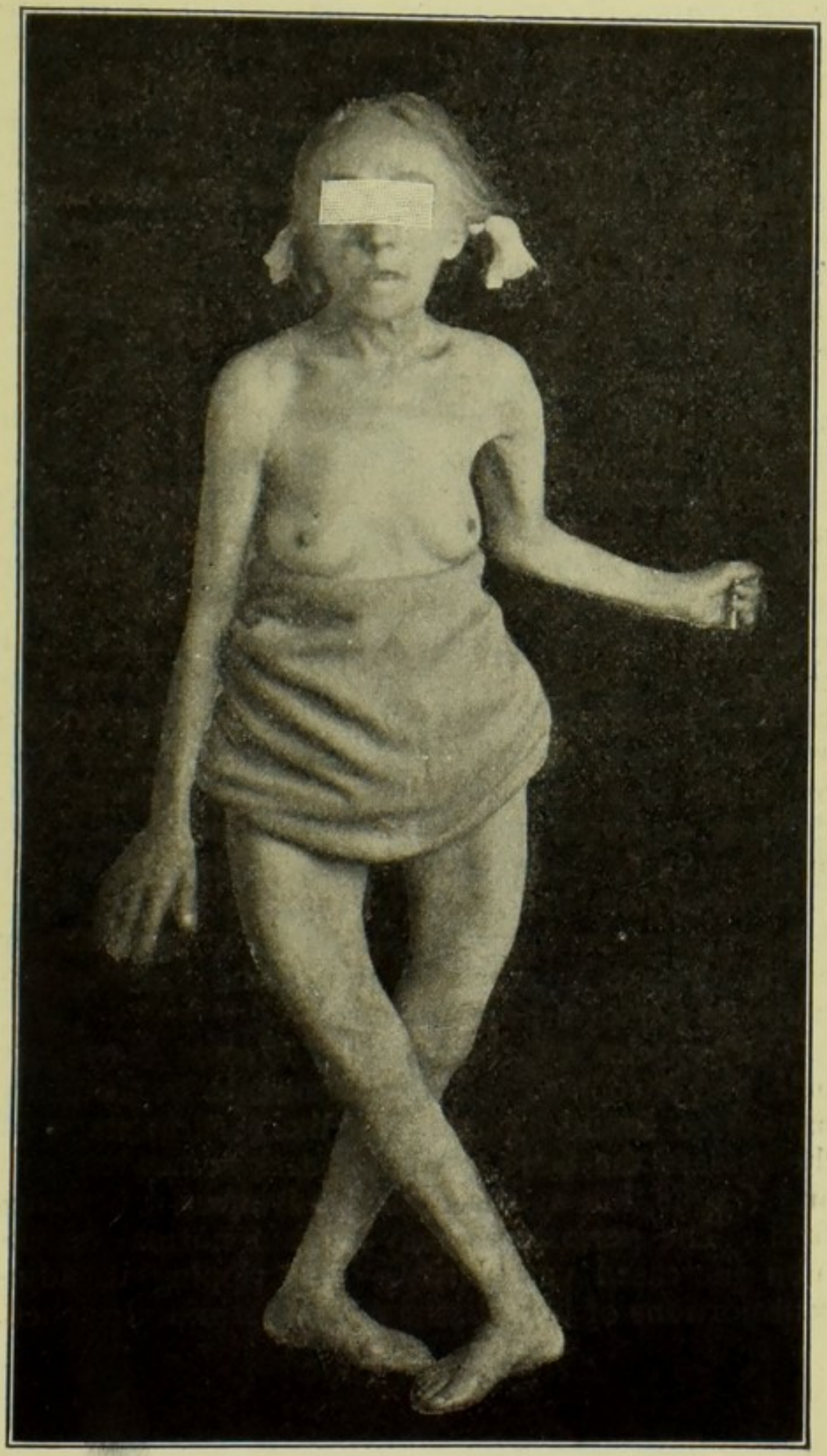

FIG. 1.—Osteitis deformans, showing bowing of legs and the only position in which standing or walking was possible.

All arguments considered, the conclusion of André Chastel, who reports two cases in his Paris Thesis of 1910, appears justified, namely, that osteitis deformans is an independent, well-defined disease, "the theory of the etiology of which has yet to be established." In the few cases in which metabolism observations have been conducted, nothing distinctive has been determined, possibly because they have not been conducted over long enough periods for so chronic a disease.

The most comprehensive study of the disease published since Sir James Paget's original account is given by Jules Vincent in his Paris Thesis of 1904, (Maladie ossieuse de Paget) in which he presents an interesting discussion of previous articles, and includes eight new cases, several of which are pictured. This writer, emphasizing the early nerve symptoms of the disease, namely the muscular cramps, fatigue pains, exaggerated reflexes, and occasional incontinence of urine and local hyperthermia, inclines to the hypothesis that the disease may originate as a central trophoneurosis, although in the few autopsies which have been made, a thorough study of possible pathological changes in the nervous system has not been undertaken. Following is a brief summary of the pathological anatomy of the disease as given by those who have opportunity to perform necropsies.

The muscles attached to the affected bones are pale and atrophic, and the ligaments occasionally have been found ossified. In the earliest stage of the disease the bone tissue appears somewhat soft and fragile, but as the disease advances it becomes firm. The periosteum may remain normal or exhibit increased vascularity. In stripping it, adjacent portions of bone tissue are sometimes detached.

The bone tissue becomes porous through enlargement of the superficial Haversian canals. The central medullary canal becomes rarefied. The sur-

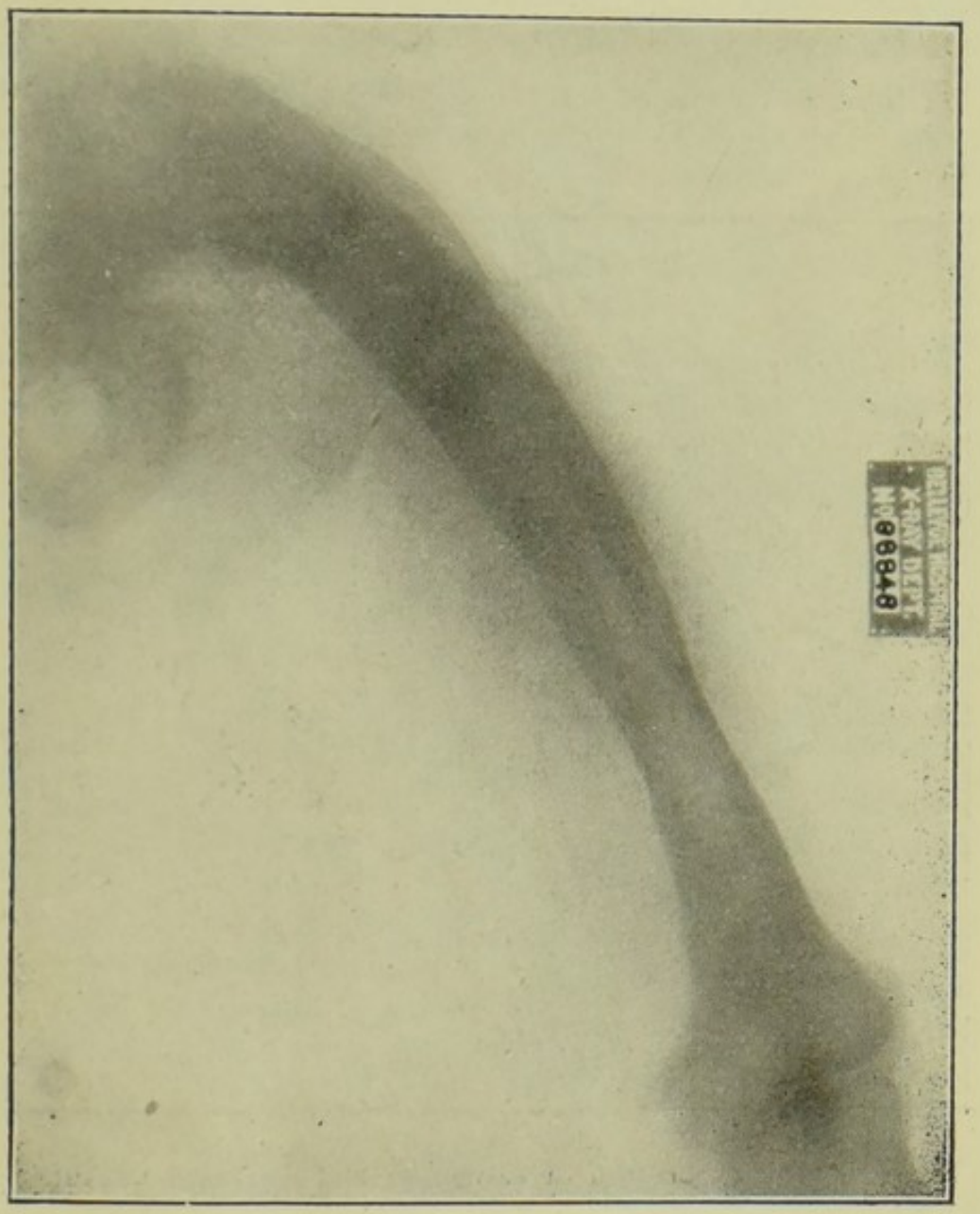

FIG. 2.—Showing bowing of left femur, hypertrophy of upper portion and rarefaction in lower third

face of the long bones is frequently uneven or mammillated, and the normal bony prominences are

exaggerated. Here and there are small areas which are so friable that they may be indented by firm pressure with the finger. The affected bone thus becomes uneven in surface and more or less nodular and spongy.

The diaphyses of the long bones become voluminous through exaggerated development of the

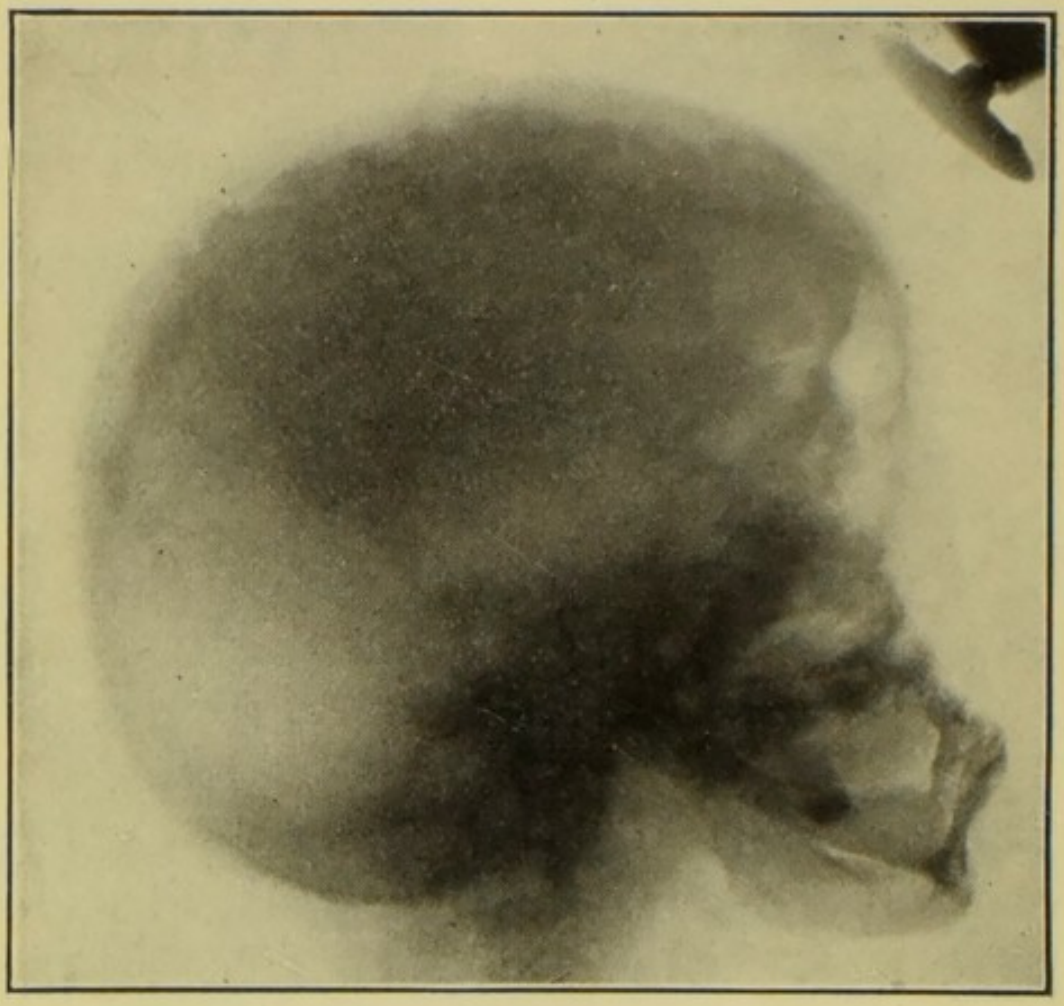

FIG. 3.—Showing breadth of calvarium and increased thickness.

compact substance although at the outer surface, and near the medullary canal, the bone is more or less rarefied. In the intermediate zone of the long bones there are islands of tissue which remain firm and solid. In parts the vascularity appears in-

creased. The medullary cavity becomes irregular in form, being here and there dilated, and elsewhere encroached upon by new bone formation. The marrow is vascular and red, resembling that of fetal bone. The spongy tissue of the epiphyses is rarefied, and at the cartilaginous surface sometimes presents fatty degeneration and necrosis.

The external surface of the cranium presents large elevated plaques. The inner surface is smooth or finely granular. The sutures are obliterated. The bones at the base of the skull are usually not thickened. On section the diploë is found to have disappeared, and the remaining tissue presents an irregular structure of alternating areas, some of which are spongy, others as dense as ivory. The ribs have occasionally been found to present rarefaction and their cartilages may be partially ossified.

Microscopically there is seen a progressive absorption of the bone tissue in the vicinity of the medullary cavity and Haversian canals, with a compensatory formative osteitis, and beneath the periosteum large porous periostoses.

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