PERIOSTITIS DEFORMANS*

BY

M. SORIANO

The Medical School, Barcelona, Spain

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In the course of 3 years we have been able to study six patients presenting similar clinical, radiological, and pathological features, which demonstrate the existence of a hyperostosing poliostotic disease, not hitherto described, which extends to the whole skeletal system except the skull, and which we have named "periostitis deformans".

Main Characteristics

(1) Clinical.—This is an acquired disease, of primary type, which recurs with varying intensity. Osseous tumours rapidly develop as a result of an intense exuberant osteogenic periostitis. Inflammatory manifestations are very rare, and if they do appear, are very discrete. The activity of these outbreaks of periostitis ceases within 2 to 12 months, and the patient then recovers his health, but the bony deformities remain as sequelae, though some of the osseous tumours may undergo total involution. These attacks may recur during the whole life of the patient, but they tend to be less intense each time. They are usually indolent in type, but are sometimes accompanied by pain that ceases once the osteophytes or affected periosteal tissues cease to grow. The more intense outbreaks are accompanied by a toxic appearance, with anorexia and wasting, which subsides once the attack has passed. There is no fever. The skin covering the bony pseudotumoral nodules becomes slightly thickened during the outbreak phase, but only rarely presents inflammatory signs. The outbreaks usually end by the

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FIG. 1.—Case 1, appearance of hands 8 months after initiation of deforming periostitis.

FIG. 2.—
Case 1,
x-ray of
hands as
in Fig. 1.

FIG. 3.—
Case 1,
x-ray of
hands one
year later.

involution of the osseous growths; a local periostitis may remain as a sequel, but there is never any suppuration, necrosis, or tumoral degeneration. The periosteal growth around the joints may cause functional loss of power which is sometimes very marked.

Many outbreaks pass off without clinical recognition and are only revealed in the radiographs which show definite superimposed layers of periostitis corresponding to the successive attacks (Fig. 5).

(2) Anatomical.—The lesions are poliostotic and frequently symmetrical; they affect the spinal column only slightly, and the skull not at all. The bones become

FIG. 4.—Case 1, Deformities of forearms and elbows during a second outbreak some months later.

FIG. 5.—X-ray of forearms as in Fig. 4. A layer of periosteal ossifying proliferation of recent appearance has developed on an old zone of periosteal enlargement remaining as a sequel of previous attacks.

surrounded by a layer of ossified thickened periosteum with exuberant exostosis of irregular form, which in very severe cases forms true bony outgrowths, causing thickening and marked deformity of the bones and peri-articular regions (Fig. 4).

(3) **Histological.**—An ossifying periosteal proliferation is observed; it first produces bone of embryonic type and this soon develops into adult bone (Fig. 6).

Almost immediately a process of osseous reabsorption begins, and abundant osteoclasts advance from below to invade the germinative layer of the periosteum, producing a spongy bone of dense mesh-work (Fig. 7). Subsequently, in this newly-formed bone, an intense osteoporosis is seen with reabsorption of the osseous tumour; this may disappear entirely (Fig. 3) though it more frequently remains as a more or less osteoporotic hyperostosis (Figs 11, 12, and 13). Before reaching the quiescent phase, the osteoblastic process, in association with the osteoclastic, takes on a mosaic-like appearance which is later replaced by an

FIG. 6.—Deforming periostitis with intense periosteal proliferation and compact bone formation (high magnification).

FIG. 7.—Periostitis in the advanced phase with spongy osseous tissue formation and a abundant osteoclastic function.

osteoporotic osseous tissue. This remains quiescent (Figs 8 and 9) until a new periosteal outbreak provokes the apposition of a new layer of osseous efflorescence, which follows the same histological evolution as the previous one.

In normal bone, an osseous condensation and eburnation is first established, and this is followed by an osteoporosis each time more marked.

The walls of the vessels that cover the periosteum present a thickening and slowly progressive hyperplasia which in advanced phases is very marked (Fig. 8).

(4) Radiological.—The x-ray photographs present four characteristic appearances:

(a) Periosteal Thickening.—This extends in the form of a layer that covers the bone with exuberant growths of capricious form.

FIG. 8.—Newly-formed osseous tissue in the quiescent terminal phase of advanced osteoporosis with enlargement of periosteal vessels.

FIG. 9.—Newly-formed osseous tissue in the terminal phase of advanced osteoporosis (high magnification).

(b) *Osteophytes or Osseous Nodules*.—These are of rapid growth; they develop on the diaphysis and in 2 to 10 months may reach the size and shape of a chick-pea or almond on the fingers, of a hazel or walnut on the forearms and elbow, and of an egg or mandarin orange on the femur.

(c) *Peri-articular Alterations*.—These develop around the joints in the form of osteophytes.

(d) *Osseous Alterations*.—At first a diffuse bony condensation is seen, and this is followed by the appearance of osteoporotic zones, larger each time, until the whole bone is invaded. In some places the cortical layer of bone underlying the hyperostosis remains striated, thickened, and porotic, suggestive of Paget's disease.

FIG. 10.—Case 2, Roentgenogram of forearm showing lesions in the spongy ossification phase initiating osteoporosis.

FIG. 11.—Case 3, Roentgenogram of femur showing large osteophytes in the advanced phase of osteoporosis.

The radiological structure of the osteophytes passes through three stages:

(i) *Embryonic Bone*.—This gives a faint radiological shadow with the appearance of an efflorescence that takes on a cockade or cauliflower shape as it grows (Fig. 2).

(ii) *Spongy Bone*.—This is seen as a sponge-like structure of dense meshes with indented or very irregular edges (Fig. 10).

(iii) *Osteoporotic Bone*.—This produces wide transparent meshes with smooth edges of capricious form.

(5) **Haematological**.—Patients so affected have a tendency to present hyperproteinaemia, the Weltmann coagulation test is shortened, and the erythrocyte sedimentation rate is occasionally raised. The serum alkaline phosphatase is always increased. Blood calcium and phosphorus are normal. There is no anaemia and the number of leucocytes is normal. Frequently a slight but definite lymphocytosis and monocytosis is observed.

Course of the Disease

Pathogenesis.—We suppose that this disease is due to a toxi-allergic action of tuberculous origin, only affecting constitutionally predisposed individuals who have in their osseous development a deficiency such as corresponds in other patients to the presentation of congenital osseous malformations. This form of *skeletal infantilism* is seen in the small size of all the patients, which is frequently combined

FIG. 12.—Case 5, Pelvis and upper half of femur showing marked residual lesions of deforming periostitis.

FIG. 13.—Case 5, Lower half of femur.

with an infantile psychopathic personality. Because of this fact of predisposition, the disease may present itself in various members of the same family.

Evolution.—The hyperplastic periostitis always appears to end favourably after a period of 4 to 12 months, but it then leaves as sequelae osseous deformities, with functional difficulties in some joints, and osteoporosis. The outbreaks may recur throughout the patient's life, though each attack is usually less intense. Though the lesions are poliostotic, in some cases they are more localized in forearms and hands, and in others in the bones of the leg.

Treatment

We have no knowledge of a curative medication of the outbreaks, nor have we been able to prevent relapses. It is possible that nitrogen mustard may be found to have a favourable action. The treatment of the sequelae by vitamin D and calcium is important, as well as the resection of any particular osseous bulgings that may hinder the movement of the limbs when present in very accentuated forms.

Summary

(1) The clinical, radiological, haematological, and histological characteristics of a type of bone deformity are described and illustrated with reference to the appearances in six patients observed during the past 3 years.

(2) The disease may be classified as hyperplastic osteogenic recurrent osteo-periostitis, of polyostotic type, with pseudotumoral exuberant development of primary nature and specific character. It is probably tuberculous in origin, but seems to require certain predisposing conditions in the skeletal development.

(3) Forms of treatment are suggested, but no method of cure has yet been found.

Périostite déformante

RÉSUMÉ

(1) On décrit et on illustre les caractères cliniques, radiologiques, hématologiques, et histologiques d'un type de déformation osseuse se rapportant à l'apparence de six malades observés pendant les trois dernières années.

(2) On peut définir cette maladie comme une ostéopériostite ostéogène hyperplasique récurrente du type polyostotique, avec un développement exubérant pseudo-tumoral de nature primaire et de caractère spécifique. Elle est probablement d'origine tuberculeuse, mais semble exiger certaines conditions prédisposantes dans le développement du squelette.

(3) On suggère des formes de traitement, mais on ne connaît pas de cure.

Periostitis deformans

SUMARIO

(1) Se describen, ilustradas, las características clínicas, radiológicas, hematológicas e histológicas de un nuevo tipo de deformidad ósea, que ha sido observada en seis pacientes durante los últimos tres años.

(2) La enfermedad puede clasificarse como una osteoperiostitis hiperplásica osteogénica de curso recurrente y de extensión poliostótica, con desarrollo exuberante pseudotumoral, de naturaleza primaria y de carácter específico. Es probable su origen toxi-tuberculoso, pero parece requerir ciertas condiciones predisponentes en el desarrollo esquelético.

(3) Se sugieren algunas formas de tratamiento, pero no se ha encontrado ningún método curativo eficaz.