OSTEITIS DEFORMANS: ITS TREATMENT BY ULTRA-VIOLET RAYS.

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OSTEITIS deformans was described for the first time by Paget in 1876, but nothing is known about its etiology. It is seldom seen before the age of 40, and is equally common in males and females. The main symptom is pain, which is attributed to rheumatism in most cases. The disease is frequently overlooked until gross deformity appears, or an x-ray photograph is taken.

Pathology.

The normal bone rarefies, and is slowly invaded by fibrous tissue, leading to softening and bending. Coincident with this destruction some regeneration and ossification occur at a slow rate. In the long bones the normal curves are exaggerated at first, and later abnormal curves make their appearance. The subperiosteal bone laid down is more porous, roughened on the surface, and considerably larger than the normal.

The long bones of the lower extremities, the pelvic bones, and the skull are most commonly affected, but any bone may be the seat of the disease. In advanced cases most bones suffer, leading to great deformity. The diagnosis is not easy when only one bone is affected.

Various treatments have been suggested, but none has proved of any value. I was prompted to write this paper by the excellent results obtained in four cases that have been under observation for nearly three

years, but the number of cases is small, and the period of observation short, for a disease progressing so slowly. The complete disappearance of symptoms, and the marked return of the bones to normality, as shown by radiograms, has tempted me to take this step with a view to getting a greater number of patients treated by this method, and having my findings confirmed or otherwise.

The cases described illustrate well the difficulties of arriving at a correct diagnosis.

CASE I.

The patient was a man, aged 55. There was nothing of importance in his family or personal history until 1925, when he was knocked down by a motor cycle and severely bruised. In 1918, however, he had complained of aching in the right groin, and was advised to diet himself. The pain diminished, but did not disappear entirely. In the summer of 1927 severe pain was felt in the same region. Sleep became impossible without hypnotics, and he was unable to follow his occupation. The local doctor suspected appendicitis, and referred him to a consultant. The diagnosis was uncertain until an x-ray photograph (Fig. 1) was taken, which showed osteitis deformans affecting the ischium.

He was seen by me in December, 1927. He looked ill owing to the pain he had suffered, the sleepless nights, and the worry of losing his employment. He was put on general ultra-violet ray treatment three times a week; at the end of twelve sessions the pain was distinctly less, and after twenty-four doses he had practically lost it. There was a marked improvement in health, and he resumed his work. With the exception of short breaks he persevered with treatment until the beginning of 1929, when a pause was suggested. After a period of five weeks there was a slight return of the aching, and treatment was restarted twice a week, when the pain again disappeared. He has since maintained excellent health in spite of a sedentary occupation and

FIG. 1.

long working hours. An x-ray photograph, taken ten months after the commencement of ultra-violet ray treatment (Fig. 2) shows a remarkable improvement in the condition of the bone.

CASE II.

An unmarried woman, aged 68, had previously been well, and there was nothing significant in her family history. In the summer of 1926 she bruised the right shin against a bath. In September of the same year aching started in the right leg, and she noticed a slight bowing. A bone-setter diagnosed arthritis, and advised exercises and diet, but the pain continued to get worse.

She was seen by me in January, 1928, when she had marked anterior bowing of the right tibia, and considerable enlargement of the diameter of the bone. A radiogram showed typical osteitis deformans, with great thinning of the compact bone. She was put on an ambulatory splint, taking the weight on the tuber ischii, in order to prevent a fracture, and a course of general ultra-violet irradiation three times a week was started. The tone of the muscles was maintained by a course of graduated faradic contractions. At the end of twenty treatments the pain had diminished considerably, and after three months she had lost all symptoms. An x-ray photograph showed regeneration of bone, and the calliper was discarded. This patient had been advised to have one-month courses of irradiation, with six-week intervals. There has been no return of symptoms, and she has maintained perfect health.

Two other cases, one in a woman, aged 54, suffering from osteitis deformans affecting the tibia, and the other in a man, aged 70, with Paget's disease affecting both femurs, have responded by disappearance of pain within three months of the commencement of treatment, and apparent arrest of the disease, as evidenced by regeneration of bone.

I much regret that the calcium content of the blood was not determined before and during the treatment; it might have thrown some light on this obscure condition.

All the cases were irradiated from a mercury vapour lamp. The area of skin exposed was increased gradually so as to raise the patients' tolerance to the rays, and the doses given were designed to produce the faintest erythema possible.

When the symptoms have disappeared it is important to impress upon the patients that treatment should not be discontinued for too long a period. This should not be difficult with a form of treatment which is not in the very least unpleasant, and is generally followed by a feeling of well-being.

PRIMARY CARCINOMA OF THE APPENDIX.

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In the last few years a considerable amount of literature in many languages has accumulated on this subject, but up to 1906 only 42 cases had been reported. By 1926 the number of recorded cases had reached 300, and last year Selinger of New York reported 34 new cases from the Post-Graduate Hospital.

Etiology.

Here there is scope for the many views as to the etiology of cancer in general. Primary carcinoma of the appendix has been reported by MacCarty and McGarth in a child of

5 years. This fact, together with the embryological and phylogenetic position of the appendix, lends force to the theory that it arises in displaced or embryonic remains; but when we ask why the displaced cells should take on malignant characteristics at this early age, we are met by the host of problems related to carcinoma in general. Again, the appendix and inflammation are closely related, and the most frequent age at which carcinoma of the appendix appears corresponds to that at which inflammation of the appendix appears. The oldest patient was aged 81 years, reported by Rogg in 1913. More cases have occurred in females than in males, but statistics here are rather confusing, because although appendicitis is acknowledged to be more frequent in males, the female abdomen is more frequently operated upon than the male abdomen, and more female appendices are sent for routine section.

The case under consideration was that of a small girl, aged 13 years, who for the previous six months had complained of occasional attacks of sharp pain in the abdomen. Recently the pain had become localized to the right iliac fossa, and was followed by nausea. The mother stated that the pains were becoming more severe and made the child cry out. The appetite was good, the bowels regular, and there was no loss of weight.

On examination the girl appeared quite healthy and well nourished. There was no anaemia, or any abnormal general features. Palpation of the abdomen detected some "guarding" of the muscles over the right iliac fossa, and there was distinct local tenderness on pressure. Per rectum nothing abnormal was detected. A diagnosis of subacute appendicitis was made, and operation decided upon.

On opening the abdomen the appendix was found lying quite free in the abdominal cavity, without adhesions. The base was thick, hard, and solid, as though a hard green pea had been inserted into it. The distal portion was slightly distended. The caecum appeared normal, and there were no enlarged mesenteric glands. The appendix was removed, together with a small cuff of the caecum, the stump infolded in the usual way, and the abdomen closed. On cutting into the nodule at the base of the appendix it was found to be very hard and solid, almost obliterating the appendix lumen.

The report on the microscopic section by one of us (H. S.) was as follows: "The sections made from the nodular appendix show a carcinoma which in places resembles the basal-celled type, and in others a scirrhous type. In most places the lining mucosal epithelium is intact, but in one place there is infiltration of the mucosa by malignant growth. The growth extends to the peritoneal surface. The stroma is mainly fibrous."

Diagnosis.

As in the case we are reporting, no one has yet made a diagnosis of primary carcinoma of the appendix before operation. It may be found post mortem, it may be found on routine microscopical examination of the appendix—which, indeed, is the reason for the increasing number of cases recently recorded—or it may be recognized when the abdomen is opened. The case presents the usual symptoms of a subacute or chronic appendicitis and clinically is diagnosed as such, or the appendix may be perforated by the growth, and symptoms of acute appendicitis ensue.

Statistics of the Mayo Clinic and other large clinics where great numbers of appendices are examined as a routine show the frequency of primary carcinoma to be under 1 per cent.

Pathology.

It has been denied that this condition is really a carcinoma, but that the tumour represents endothelial proliferation as a result of inflammation or displaced epithelial cells. Hertzler points out in support of this view that the usual round-cell infiltration is absent, and that the connective tissue retains its acidophilic properties. For this reason the name "canceroid" has been suggested. Fortunately, the condition is generally benign, but there are several records of metastases in the liver, the peritoneum, or the lymph nodes. Also, although the growth is generally confined to the mucous and submucous layers with a tendency to infiltrate the muscular layer, it is nearly always limited by the serosa. Different types of cells have been found. The spheroidal-celled type is most common, but, as one would expect, cuboidal, cylindrical, endothelial, and mixed types and colloid degeneration may all be represented.

Selinger, in his article in the *Annals of Surgery* (1929), quotes the latest word by P. Masson:

"Carcinoids result from proliferation of the intranervous argentaffin cells of the neurocoelomic type. They pile up in the nerve fibres, finally rupture their sheaths, and infiltrate the intestinal tissue of the mucosa and then that of the submucosa."

Conclusions.

Primary carcinoma of the appendix is not so rare as to constitute a pathological anomaly, but is becoming more and more widely recognized. Occurring as it may do in young children, it provides a further field for research and speculation on the origin and nature of cancer in general. Considerations of the phylogenetic and ontogenetic development of the appendix together with this pathological change may throw further light on the problem.

The general benignity of the growth, unless it occur at the base of the appendix or infiltrate the neighbouring structures, suggests that treatment should be limited to removal of the appendix without resection of the ileo-caecal region.

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SYMMETRICAL GANGRENE COMPLICATING RHEUMATOID ARTHRITIS.

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RAYNAUD's disease has been reported from time to time as a complication of rheumatoid arthritis, giving rise to attacks of local syncope and asphyxia in the extremities, and loss of tissue, notably in the ears and the tips of the fingers. Llewelyn Jones mentions that in one advanced case in which Raynaud's symptoms developed, areas of skin over both heels underwent a necrotic change. Records of cases of symmetrical gangrene due to other causes are difficult to find, and it is for this reason that the following case was thought to be of interest.

A married woman, aged 38, with two children, stated that as a child she had always had cold hands and feet. Eight years pre-

Fig. 1.

viously she was first taken ill with an attack of acute rheumatism affecting both feet, the left ankle, both knees, and both hands and wrist-joints. The condition became gradually worse, treatment at Bath affording little relief. Six weeks before admission to hospital she noticed that her hands and feet were very cold and painful, and that the circulation was poor. Four weeks later she discovered that the right foot and the left hand were changing in colour, gradually becoming black; this change was accompanied by excruciating pain, more especially at night.

The patient was admitted into hospital on November 18th, 1929. She was pale and markedly wasted, and all four extremities showed advanced arthritic changes of the rheumatoid type. There was well-marked dry gangrene of the right foot affecting all the toes and extending proximally for two inches along the dorsum of the foot, with a well-marked line of demarcation; there was also gangrene of the left hand (see photograph, Fig. 1). The left foot