OLLIER'S DISEASE: DYSCHONDROPLASIA

BY

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The introduction of Roentgen rays enabled Ollier in 1899 to describe the disease now generally known by his name. He summarized it as "an affection characterized by irregularity in development, evolution, and distribution of cartilaginous tissue in the bones in process of development, the cartilage seeming to be of normal structure and changing more or less in the direction of ossification." Put more clearly, we may say that Ollier's disease, dyschondroplasia, is characterized by the presence of persistent areas of cartilage in the diaphyses of long bones and short long bones as a result of the failure of normal ossification of cartilage to take place.

Ollier's dyschondroplasia is now clearly differentiated from Keith's diaphysal aclasis, a disease having as its main features multiple exostoses with a strong hereditary factor. Sir Arthur Keith (1920) showed that the so-called exostoses in diaphysal aclasis were in reality bony outgrowths due to defective tubulation in the remodelling process of the

FIG. 1.—Amputation stump and ulcers in the groin, showing the thick everted edges.

long bones during growth. In Ollier's disease there are gradations from persistent islands of cartilage in the metaphysis and diaphysis seen only in the radiographs (and here the importance of radiographs of the whole skeleton is emphasized) to large palpable masses of cartilage. In addition there are small exostoses of irregular formation, not pedunculated and not characteristically pointing away from the end of the bone as are those in diaphysal aclasis. It was partly the presence of these exostoses which has caused confusion in the literature between Ollier's disease and Keith's diaphysal aclasis, but, in addition, it is evident that there are cases occupying an intermediate position.

The failure of normal ossification of cartilage to take place in the growing ends of bone has as its result that there remain behind in the diaphysis areas of cartilage which do not ossify. These may not be discernible clinically or there may be palpable masses of cartilage to which the term "tumour" in the broadest sense of a lump may be applied. This abnormal growth is not, then, primarily one of true tumour formation, but it is evident that the distinction between a mass of cartilage due to abnormal growth and a true chondroma or neoplasm of

FIG. 2.—The hands.

cartilage will be a narrow one. The fact that these masses are actually cartilage has been well established by biopsy.

Certain additional features in the case here reported clearly identify it as belonging to the Maffucci syndrome. This consists of the characteristic features of dyschondroplasia, mostly on one side of the body, with haemangiomas more or less confined to the opposite side. Malignant changes have been observed in some cases in the literature, and they also supervened in the following case.

Case Report

The patient, a man aged 34, was admitted to Whipps Cross Hospital on January 14, 1948. He was apparently normal at birth. Swellings of the left hand were first observed at the age of 5, after his hand had been caught in a door. Later on a nodule came up on the right hand, after it had been struck by a cricket ball. He said he had fractured his left leg eight times while playing football, the last time being in 1930. There was no family history of similar disease.

FIG. 3.—Radiograph of the left hand. The nodular masses consist of cartilage with areas of calcification.

FIG. 4.—The left foot, showing multiple hard nodules of the phalanges.

I am much indebted to Dr. H. G. Close and Dr. Malcolm Pearson for the following notes taken after his admission to the Southern Hospital, Dartford, Kent, on February 12, 1945. He was then complaining of pain and a swelling of the right ankle and foot. The right leg was found to be 1 ft. (30 cm.) longer than the left. On the outer side of the right foot was a hard spherical mass with ulceration of the skin below. Biopsy showed this to be a lymphangioma without definite evidence of malignancy. On September 4, 1945, a right below-knee amputation was performed. The skin flaps did not heal satisfactorily, and in January, 1946, enlarged glands were palpable in the right groin. On June 5, 1946, a mid-thigh amputation was performed with the flaps left open, with subsequent secondary suture. On August 14, 1946, inguinal lymph nodes were removed and reported on by Dr. A. H. T. Robb Smith as a "malignant lymphangioma." He subsequently returned home and worked on a barge until Christmas, 1947. The amputation stump then broke down and he developed an ulcer of the right groin.

On admission to Whipps Cross Hospital his condition was as follows. The amputation stump of the right thigh was the seat of a malignant ulcer some 6 in. (15 cm.) in diameter with a thick everted edge and a sloughing floor. There was profuse foul-smelling discharge. In the groin were two

FIG. 5.—Radiograph of the pelvis, showing extensive chondromatosis of both wings of the ilia.

ulcers with similar thick everted edges (Fig. 1): a large lower one 4 in. (10 cm.) in diameter over Scarpa's triangle and an upper smaller ulcer near the symphysis pubis. The healed biopsy incision can be seen in the photograph, the ulcers evidently being due to fungation of further malignant glands in the groin.

The hands are seen in Fig. 2. The fingers and thumb of the left hand show hard, irregular, but fairly smooth masses over which the skin is tightly stretched. Blue veins course over the cartilaginous masses. He was able to move his fingers a little and some nails were still present. Sensation was present. The right hand showed chondromata of the thumb and little finger only, and was a useful member.

The radiograph of the left hand (Fig. 3) shows the nodular masses on the fingers to be cartilage with areas of spotty calcification. The

phalanges and the interphalangeal joints have almost disappeared, but a thin layer of cortical bone remains in some places. The metacarpals approach more nearly to normal, but they are shortened and there are obvious areas of cartilage with spotty calcification in the epiphyses, metaphyses, and diaphyses. The carpus is practically normal. The lower ends of the radius and ulna are broadened and somewhat striated, and there are areas of cartilage and spots of calcification. No Madelung's deformity is present.

Fig. 2 shows the Madelung deformity of the right hand, and a radiograph showed the shortened broad radius with some persistent cartilaginous areas, although the lower end of the ulna is not much affected and does not illustrate the bayonet shape mentioned in the literature. There are irregular cartilaginous and bony outgrowths of both phalanges of the right thumb, and a nodule of cartilage in the head of the metacarpal. Two areas of calcification are seen in the head of the second metacarpal, but otherwise the metacarpals and phalanges of the index, middle, and ring fingers are normal. The little finger shows areas of cartilage and bony outgrowths in the distal end of the metacarpal and all phalanges. The carpus is normal.

The left foot (Fig. 4) showed multiple hard nodules of the phalanges, especially of the second, third, and fourth toes, with the remainder of the foot normal. A radiograph showed the tarsus to be more or less normal, though the astragalus has some deformity of its articular surface. There is a clear area in the os calcis near the calcaneo-cuboid joint. Broadening and striation of the lower end of the tibia and short irregular bony outgrowths are present. The masses in the toes are translucent and largely cartilaginous.

The face and skull were normal and showed no asymmetry. The sclerotics were of normal colour. The patient was of

FIG. 6.—Radiograph showing the short left femur, the lower end of which is seen to be greatly broadened.

good intelligence and stated that he had been getting around on crutches and working on barges until shortly before admission to hospital. A radiograph of the skull was normal. There were no patches of pigmentation on the skin. The cardiovascular system and lungs were normal clinically. The urine was normal—no Bence-Jones proteose was present.

The abdomen was normal to palpation, but a radiograph of the pelvis (Fig. 5) showed extensive chondromatosis of both wings of the ilia. These were not abnormal to palpation. The absence of dense radiating lines in the ilia (raies sombres of Voorhoeve) is noteworthy. As pointed out by Hunter and Wiles (1935), this is probably due to the age of the patient, the radiating lines appearing in the pelvis and other parts of the skeleton in children only.

An x-ray film showed a little broadening of some ribs, but no evidence at that time of metastases in the lungs.

The left thigh was short for the patient's general build, and hard masses were felt at the lower end of the femur and upper end of the left tibia and fibula. He had a good range of movement in the knee-joint. Fig. 6 shows the rather short left thigh, but the lower end of the femur is particularly interesting. The lower end broadens in the trumpet-like fashion of diaphysal aclasis, with a broad metaphysis, its sides being approximately parallel. There are extensive areas of cartilage persisting in the metaphysis and epiphysis, and some deformity is evident in the lateral view (Fig. 7). The areas of cartilage in the tibia and the irregular small exostoses, not pointing towards the ends of the bone, are seen.

The angular bends of dyschondroplasia described by Jansen are well seen in the radiograph of the humerus (Fig. 8). The shoulder- and elbow-joints are normal, though the upper ends of the humerus are somewhat broadened. There is a spindle-shaped swelling of well-calcified bone in the middle of each humerus with angulation. There has evidently been dysplasia earlier in growth, with "healing" as Ollier described. Little persistent cartilage is here seen in the shafts.

A blood count showed: haemoglobin, 76%; red cells, 5,100,000; colour index, 0.74; mean diameter of red cells, 7.3 μ ; white cells, 13,500 c.mm. (polymorphs 79%, lymphocytes 10%, monocytes 5%, eosinophils 6%). Serum calcium was 10 mg. per 100 ml.; plasma phosphate 2.3 mg. per 100 ml.; serum acid phosphatase 3.2 units; serum alkaline phosphatase 5.5 units. The Wassermann and Kahn reactions were negative. Biopsy of the amputation stump showed malignant lymphangioma.

Course.—Deep x-ray therapy at the London Hospital under Dr. Frank Ellis caused considerable improvement in the fungating ulcers of the amputation stump and groin. The ulcers

cleaned up, the thick everted edge became flat, and some epithelization from the edges took place. Subsequently, however, his condition deteriorated and metastases appeared in the lungs. He died in November, 1949, and permission for necropsy was not obtained.

Comment

This case illustrates the essential features of Ollier's disease recorded in the considerable literature on the subject, and the history and appearances may be regarded as typical. The prognosis as regards life appears fairly good, considering that the Maffucci syndrome must be regarded as a mesodermic dysplasia beginning in foetal life. Our patient was 35 years of age at death, and of the 20 cases traced by Carleton *et al.* (1942) the average age was 33. The high proportion of cases (25%) which became malignant is noteworthy.

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FIG. 7.—Lateral view of left femur and tibia, showing areas of cartilage in the tibia and irregular small exostoses.

FIG. 8.—Angular bends of dyschondroplasia in the humerus.