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DYSCHONDROPLASIA*

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Dyschondroplasia has been definitely removed from the category of tumors and is now recognized as a disturbance of bone growth which begins in childhood. It is characterized by the expansion of the ends of the long bones and by the appearance of multiple cartilaginous growths at the ends of those bones.

This disorder is not an uncommon one, for in reviewing the literature on this subject there are to be found about 400 articles dealing with over 700 cases. These include cases drawn from the literature of all countries; the following are the percentages from the various countries:—Germany, sixty per cent.; France, twenty-seven per cent.; England, eight per cent., other countries five per cent. Thus this disorder appears to be world-wide in its distribution, Germany having the greatest number of recorded cases.

Cases showing the typical characteristics of this disorder and the influence of heredity have recently been described from India and other parts of the Far East. Taking the recorded cases as a whole, males appear to be affected more than females, approximately in the proportion of three to one; but in the limited number of cases reported from India, the females slightly predominate.

The great majority of cases are associated with stunted growth; the stunting is confined to the limbs only, the trunk maintaining its normal length, but a few cases of gigantism with multiple exostoses have been described in the French literature.

Heredity plays a very important part in this disorder; this was clearly brought out by Gcassage and Carling in an exhaustive paper on this subject read before the Royal Society of Medicine in 1910. These writers demonstrated that it frequently extends throughout several generations, and that it is rare to find isolated cases; in these so called isolated cases one generally finds on investigation quite an extensive family tree, but occasionally a generation may be skipped. The line of descent appears to be distinctly more paternal than maternal; and there is no case on record in which transmission has taken place through an unaffected male, although the malady may be passed on by an unaffected female. Many family trees have been worked out more or less completely.

The appended tree is that of a Chinese aged fifty-two, the details of whose history are recorded later in this paper. This family tree is for the male members of the family only. Little or no records were

*Read before the Surgical Section of the Far Eastern Association of Tropical Medicine, at Bangkok, Siam, December 8, 1930.

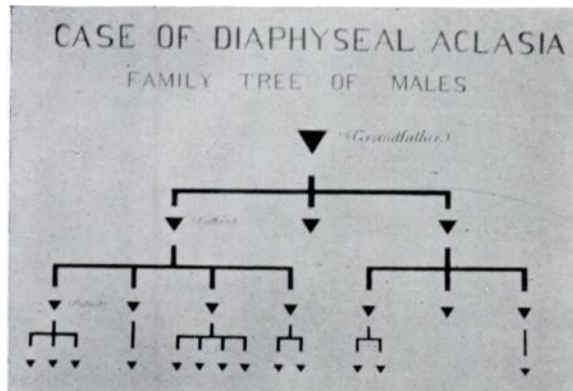


FIG. 1

Family tree of four generations showing the affected males only.

obtainable regarding the female side of the family as the daughters of Chinese do not seem to receive the same consideration as do the sons. The father in this case has had children by two Siamese wives and the children by both wives are affected, indicating very clearly that the transmission is definitely paternal.

CASE REPORT

The following case illustrates well the family tree and malignant changes developing in an exostosis. The patient is a Chinese aged fifty-six years, an opium addict, who came to the Siriraj Hospital, Bangkok, on July 12, 1929, complaining of a swelling of the left knee and left foot which prevented him from walking.



FIG. 2

Patient, showing size of tumor.

On investigation he gave a family history of the same malady affecting his grandfather, father, brothers, children, and nephews. The family tree is shown on the accompanying chart.

His present illness dates back to childhood when he noticed a hard mass on each thigh just above the knee, more marked on the inner side than the outer. These masses slowly increased in size until the age of twenty-one, then they remained stationary, not causing any trouble until the age of forty. Fifteen years ago, the left one slowly began to increase in size; this slow enlargement continued until six months ago, when the rate of growth became much more rapid; so that now he is unable to walk from sheer size of the tumor. Although the tumor has increased in size, he has felt fairly well, but he says that lately he has lost weight.

Examination:—The accompanying photograph gives the best idea of the size of the tumor. In the right popliteal space there is a large pointed exostosis and also one on the right scapula which is causing considerable depression on the ribs on that side.

Laboratory examinations:—

Blood:—Red blood cells—3,090,000

Hemoglobin—50 per cent.

White blood cells—7,300

Color index—.83

Wassermann reaction—negative

Kahn test—2+

Urine:—Trace of albumen with hyaline and granular casts.

X-ray Examination:—Film of the left thigh and knee shows an enormous tumor formation involving the lower two-thirds of the femur. On the posterior surface of the femur there is a remnant of the exostosis from which the tumor arose; small irregular islands of ossification are scattered throughout the mass. Film of the right femur shows a large pointed exostosis. The chest shows no evidence of metastases.

Diagnosis:—Dyschondroplasia with sarcomatous degeneration, arising from an exostosis on the femur.

Treatment:—Amputation in the region of the left hip joint was advised; to this the patient readily consented. The operation was performed on July 21, 1929, under spinal anaesthesia; hemorrhage was controlled by a tourniquet applied above two Wyeth's pins. An external racket incision was made and the femur was divided just below the lesser trochanter. The patient made an uninterrupted recovery.

Pathological examinations:—The specimen is the complete leg with a huge lobulated tumor involving the lower two-thirds of the thigh and weighing 19.75 kilograms. On section the growth proves to be composed of grayish translucent cartilaginous tissue which involves the epiphysis of the lower end of the femur and extends upwards in the marrow to just below the section of the bone at the line of operation. It has perforated the compact bone four centimeters above the epiphysis and extends outwards in all directions. The tumor tissue itself is completely enclosed in a thick layer of grayish firm tissue and contains scattered irregular spaces filled with blood-stained fluid.

Microscopic section of the growth shows it to be made up of irregular masses of closely packed cartilaginous cells intersected by bands of fibrous tissue in which are blood vessels. The tumor cells have many mitotic figures and in general have undergone mucinous degeneration; in a few places they show calcification.

Pathological diagnosis:—Chondrosarcoma.

CONCLUSIONS

1. There is described a case of dyschondroplasia occurring in a Chinese, showing a family tree well marked with the disease and malignant degeneration occurring in one of the exostoses.

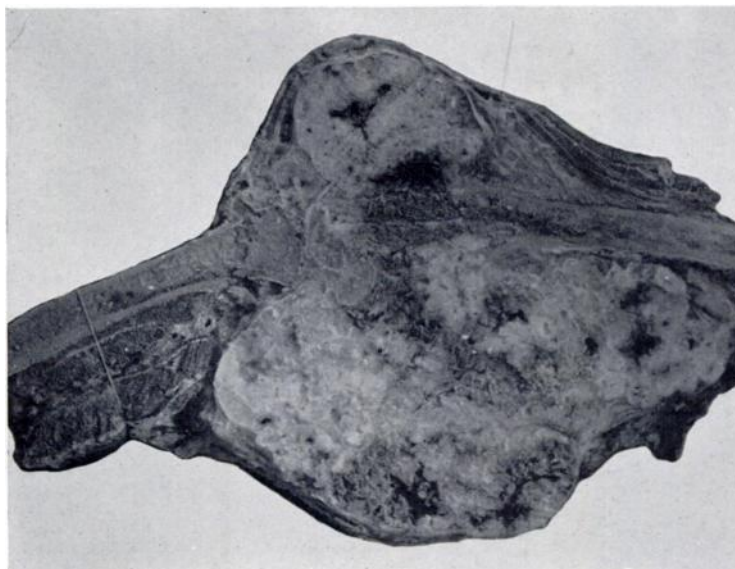


FIG. 3
Section of amputated limb.

2. In any case of dyschondroplasia in which an increase in the size of an exostosis takes place after growth of the skeleton is complete, it is strongly suggestive of malignant degeneration at that point.

3. Dyschondroplasia has a world-wide distribution and in the East it shows the identical characteristics that it does in the West.

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