LOCAL GIGANTISM

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Local gigantism is uncommon. Kanavel (1932) asserted that less than seventy-five cases of megalodactyly had then been reported. Referring to this condition Elkeles (1951) stated: “Recording of rare congenital disorders is not only justified on scientific grounds, but their study and grouping may throw light on problems of physiology and on the etiology of diseases.”

CASE REPORT

The patient was a European girl of two and a half years. There was no family history of congenital deformity. She was an only child. Her past illnesses included an attack of cerebral malaria at the age of thirteen months, a second attack of malaria at the age of seventeen months, and an attack of acute tonsillitis at the age of sixteen months. She had otherwise been well.

When the child was two and a half years old her mother first noticed, while bathing her, that she had enlargement of her left thumb and right little finger. She had apparently suffered no pain and had not been feverish. Examination revealed diffuse fusiform swelling of the left thumb in the region of the interphalangeal joint, and of the right little finger at the level of the proximal interphalangeal joint (Figs. 1 and 2). There was no reddening of the skin or increased heat over the affected parts. There was no tenderness or limitation of movement. There was no affection of any other bone or joint.

Further examination revealed no abnormality of heart, lungs, abdomen or nervous system. No nodules were found in skin or subcutaneous tissue. No areas of pigmentation
were seen on the skin. There was no thickening of any peripheral nerves. There was no enlargement of spleen, liver or lymphatic glands. Her teeth and tonsils appeared healthy. The child had a healthy general physical appearance.

*Investigations*—Leucocyte count showed total leucocytes 13,500 per cubic millimetre (neutrophils 42 per cent, lymphocytes 58 per cent). No abnormality of the cells was seen. Urine examination revealed no abnormality. The Kahn test was negative. The Mantoux test (1 in 1,000) was negative. The erythrocyte sedimentation rate was 5 millimetres in 1 hour (Westergren method).

*Progress*—The child was observed at intervals for three and a half years. At the age of five and a half years enlargement of the right second and third toes was observed. The left ring finger had also become slightly swollen. Examination six months later showed enlargement of the left thumb, left ring finger, right little finger and right second and third toes. The left thumb and right little finger had remained almost stationary over the past year, though measurements suggested that the thumb had decreased very slightly in circumference.

*Fig. 2*  
Photograph showing marked enlargement of the left thumb.

*Radiographic examination* (Figs. 3 and 4)—The proximal and middle phalanges of the right little finger, the metacarpal, proximal and distal phalanges of the left thumb, and the proximal and middle phalanges of the left ring finger showed hypertrophy of their diaphyses without evidence of a disease process. Similar but slighter changes were seen in the right second and third toes. The appearances suggested congenital local hyperplasia (local gigantism) of a mild type. Radiographs of the lungs revealed no abnormality.

**DISCUSSION**

The etiology of local gigantism is obscure. The deformity has been congenital in several of the reported cases (two cases described by Elkeles (1951), four cases of Moore (1942), one case of Inglis (1950), one case of Capel (1935) and one case of Kaplan (1947)), but Capel (1935) put forward the following arguments against its being a purely genetic condition: "In only one of the reported cases is there a family history of the condition: in almost every case it is unilateral, and only rarely is it associated with other congenital abnormalities. The other congenital abnormalities of the fingers which do give clear evidence of inheritance are usually bilateral, and they are very frequently associated with other congenital defects." Cases of
FIG. 3
Radiographs of hands.

FIG. 4
Radiographs of feet.
bilateral but asymmetrical macrodactyly have, however, been reported by Killinger (quoted by Kanavel 1932), by Mouriquand and Buche (quoted by Kanavel 1932) and by Capel (1935). Moore (1942) described bilateral local giantism of toes. The case reported here shows a combination of local giantism of both hands with a similar condition of one foot. The lack of symmetry between the two sides in most of the recorded bilateral cases is of significance.

As Moore (1941) pointed out, local giantism is unlikely to result from the action of a hormone, "which by its very nature must have access to all parts of the body."

Weber (1918, 1951) divided local hypertrophy into two categories: 1) true giantism, in which the blood vessels are not more involved than the other tissues of the hypertrophied part; and 2) angiectatic hypertrophy. Wright (1951) considered that haemangiomatous and lymphangiomatous conditions have much in common, and that it is possible that they both cause alteration in the size of the developing limb, sometimes an increase, sometimes a decrease. One of Elkeles's (1951) patients displayed a lymphangiomatous condition of the right palm in association with local giantism of the right second and third fingers.

Evidence strongly suggests that macrodactyly is related to neurofibromatosis (Inglis 1950). Brooks and Lehman (1924) stated: "We know of no other condition in which there is a spontaneous excessive growth in length of a single bone other than Von Recklinghausen's disease." The presence of neurofibromatosis may, however, be overlooked in children, for Moore (1942) noted that cutaneous tumours seldom occur in children, even when typical pigmentary changes are present; but he stated that examination of the finer terminal branches in cases of proved neurofibromatosis showed a pathological picture strikingly like that in the sections from cases of macrodactyly. Cases of local giantism associated with neurofibromatosis are recorded by Moore (1941, 1942), Bell and Inglis (1925), Baraldi and Ruiz (1934) and Brooks and Lehman (1924), who give references to other cases. Cases associated with areas of pigmentation, but without cutaneous tumours, are cited by Elkeles (1951), Moore (1942), Adair, Pack and Farrior (1932) and Kaplan (1947). A family history of Von Recklinghausen's disease, which was regarded by Moore (1942) as collateral evidence since it is frequently hereditary, was recorded by him in one of his cases of local giantism. Other conditions that have been shown by Adair et al. (1932) to have close clinical analogy to neurofibromatosis and which have been found in cases of local giantism are multiple lipomata (Inglis 1950, Geunther quoted by Zondek 1944, Moore 1942 and Adair et al. 1932) and angiomata (Moore 1942, Elkeles 1951).

As Moore (1941) pointed out, the strongest argument in favour of the view that the neurofibroma is the cause of the hypertrophy is the fact that there is a definite segmental relationship between the affected nerve and the overgrowth, as was well demonstrated by his own case of marked fusiform enlargement of the right median nerve associated with giantism of the right two middle fingers, and by that of Bell and Inglis (1925) of plexiform neuroma of the median nerve leading to the enlarged part. A similar segmental relationship between neurofibromatosis and splanchnomegaly has been observed by Winestine (1924), who recorded a case presenting papillary adenomatosis of the rectum combined with neurofibromatosis of the pelvic and mesenteric plexus, and who referred to accounts by Oberndorfer (1921), Schultz (1922) and Schmincke (1922) of giant appendices, and by Pick (1923) of circumscribed overgrowth of a segment of intestine, associated with neurofibromatosis of their nerve supplies.

Moore (1942) pointed out that it is generally accepted that neurofibromatosis is a congenital condition, and that it passes through various phases of development. The increase in endoneural fibrous tissue found by him in cases of macrodactyly in children may be an early stage in the development of neurofibromatosis at a later age. The difference in the rate of development of neurofibromatosis (Ford 1952) would account for the fact that some cases of local giantism are congenital, whereas others, such as that described here, appear later. The irregular distribution and usual lack of symmetry in bilateral cases are explained by the variable distribution of neurofibromata.
Different views have been put forward to explain the association between neurofibromatosis and local gigantism. Brooks and Lehman (1924) considered that the overgrowth results from invasion of the bone by the neurofibroma, explaining the cases of bone shortening by destruction of epiphyseal cartilage. But Moore (1941) stated: "We find it difficult to accept this theory that the bone grows throughout its length, since it runs so directly counter to all recent work on the growth of bone. Furthermore, we can find no report of actual invasion of the shaft by the neurofibroma... We believe that overgrowth takes place at the epiphyseal line as in normal bone, but at a greatly accelerated rate." It should, however, be noted that Kaplan (1947) described a patient aged twenty-four whose right thumb had become larger during the past two years. Moore (1941) considered that the bony abnormalities result from the changes in the nerves. He stated: "Certainly in these cases there is a defective pattern of growth associated with a defective nerve." He believed that the mixed over-development and under-development that he found were the outcome of faulty control over growth. He found evidence of endarteritis and stated: "Possibly the autonomic nervous system which controls the arteries may also have some control over growth."

Inglis (1950) considered that the elongation of a limb associated with osteochondromata (recorded in two cases by himself and in one case by Bell and Inglis (1925)) and with haemangiomata (described by Bell and Inglis 1925) in an area of neurofibromatous involvement result from neural intrinsic factor of the neurofibromatosis. On this supposition, presumably not only angiomata but also multiple lipomata, which are recorded in association with neurofibromatosis, would have a similar origin.

SUMMARY

1. A case of local gigantism is described, with enlargement of the left thumb, the left ring finger, the right little finger, and the right second and third toes.
2. No other record can be found in the literature of an association of local gigantism of fingers of both hands with a similar condition of the toes of one foot, though there is no theoretical reason why macrodactyly should not affect the digits in any combination.
3. Theories regarding the etiology are discussed.

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