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MEGALODACTYLISM

Report of 7 Cases

By

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Megalodactylism is a rare and peculiar disease consisting in a grotesque overgrowth of one or more fingers. The overgrowth is present at birth and increases somewhat during childhood, but usually stops at puberty. The abnormal growth comprises all tissues, bones as well as soft tissues, and always appears to be associated with considerable thickening, or even tumor formation, of the median or ulnar nerve and their branches.

Apparently, the increased growth is maximal along an axis, decreasing to both sides of it. This lends the digits a lateral curvature away from the axis (7). As a rule only one or a few fingers are affected, while the others are entirely normal.

The aetiology of the disease is unknow, and there are no prophylactic measures. Therefore, the treatment has to be restricted to amputations and plastic operations, thinning out the hypertrophic soft tissues. In addition, some cases have been treated by epiphysiodesis (3, 6), which has proved a successful check on longitudinal growth, but of course has not reduced the thickness of the giant fingers.

At times, there is such pronounced thickening of the median nerve that this has required operation with removal of the tumour, possibly together with the nerve which is in some cases 1 cm in thickness; the digital nerves may be as thick as a lead pencil. If the nerve is traced proximally it gradually approaches normal dimensions.

The microscopic appearances of the removed tissue are uncharacteristic. As is apparent from the following case reports, the histological diagnoses are "lipoma", "fibrolipoma", or "lipo-fibroneuroma". The appearances are reminiscent of those described by Dejerine-Sottas

as hypertrophic neuritis, which, however, is not associated with gigantism. No signs of malignancy have been found.

Other designations of this rare disease also refer to the nerve changes: "elephantiasis nervorum" and "von Recklinghausen type of macrodactylism" (11). However, although the microscopic picture may at times remind of von Recklinghausen's neurofibromatosis, the patients usually do not exhibit the subepidermal neurofibromas characteristic of this disease.

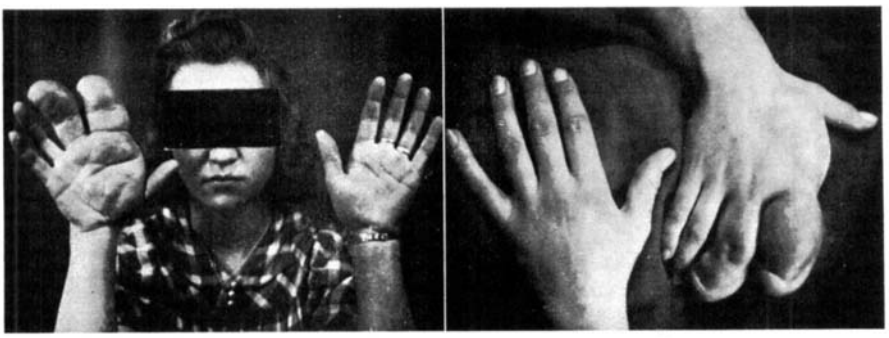
The disease has also been called "macrodystrophia lipomatosa" because of the very ample soft tissues on the fingers and adjacent parts of the hand (8).

The few previous cases have been published mostly one by one (3, 6, 8, 9, 10). In the Departement of Hand Surgery of the Orthopaedic Hospital, Copenhagen, we have collected 6 cases in the course of the past years which will be reported below together with one case affecting the foot. Some authors (6) have stated that the overgrowth most often affect the long and ring fingers, while others say that the index and long fingers are most often involved (1, 7). The present 6 cases confirm the latter view, as the *index finger* was involved in all 6 cases, the *long finger* in 4, the *thumb* in 2, and the *ring finger* in only 1 case. The little finger was not affected in any case.

Arterio-venous anastomoses or aneurysms may give rise to some overgrowth, but hardly of this special nature. Arteriography, done in Case 3, showed no vascular abnormalities.

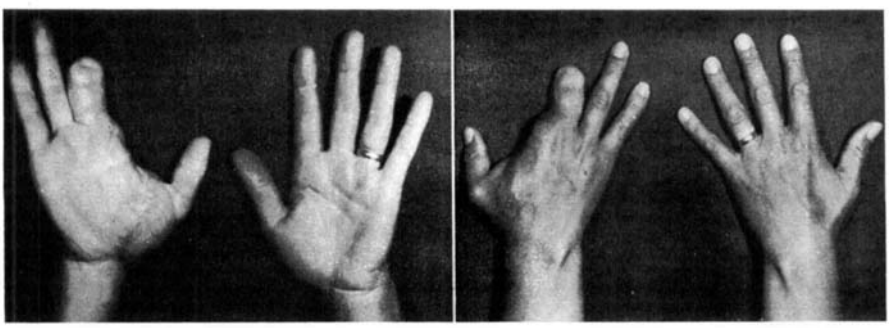
No hereditary features were demonstrated. Among the patients' closest relatives there had not been deformities other than varus feet (Case 3). As to some action which may have been operative during foetal life we can only surmise. Regrettably, it was impossible to study this aspect in the reported cases, mainly because the patients were born long before attention was directed at toxic actions during pregnancy. On the basis of the knowledge about the earliest embryological development (4, 12) and in analogy with the contemplations concerning other extremity malformations (5), it is reasonable to believe that the cause may have been embryological or foetal.

Giant growth may occur also in sites other than the fingers. As in Case 7 it may involve the toes (10), and it may affect an entire limb (2), but the literature also includes description of cases with hemilateral giant growth and hypertrophy of the contralateral cerebral hemisphere (2). In principle, this "hemihypertrophy" is similar to the local changes described in the present paper.



a *b*

Figures 1 a and b: Case 1, female, aged 16.



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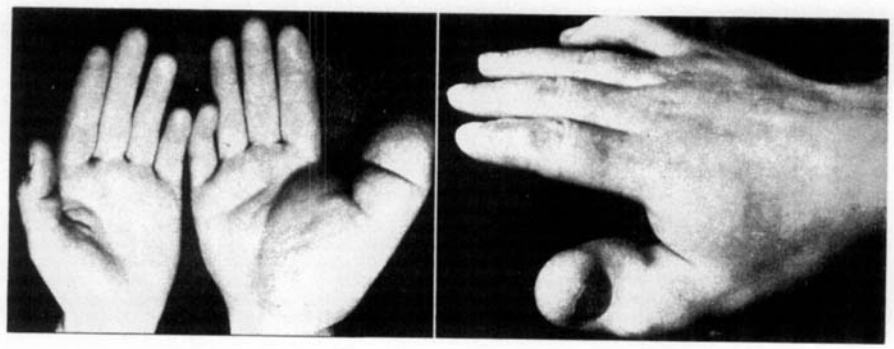
Figures 2 a and b: Case 1, at 38 years of age.

Of the 7 patients (the case affecting the toes included) 3 were females and 4 males. All the cases were unilateral.

Case 1.

A woman, now aged 38, with giant growth of the index and long fingers from birth. At the age of 4 years the distal phalanx of both these fingers had been removed at the local hospital. She was referred to the Department of Hand Surgery because of thickening of the soft tissues on the volar aspect of the hand from the wrist to the tips of the index and long fingers and considerable giantism of these fingers (Figures 1 a and b).

At the age of 16 she had an operation removing the greater part of the ample soft tissue, a total of 155 g. Grossly, the specimen looked like typical lipoma. Later, part of the middle phalanges of both fingers was removed. Thereafter, the condition remained stationary for some years, but then she started to have increasing complaints due to increased growth of the soft tissue, mainly in the web space between



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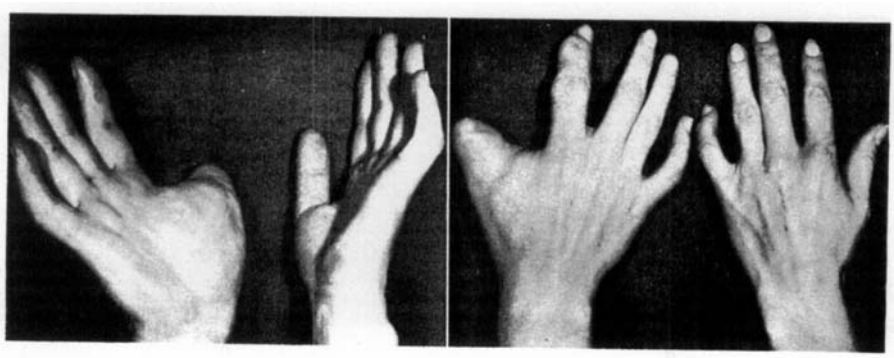
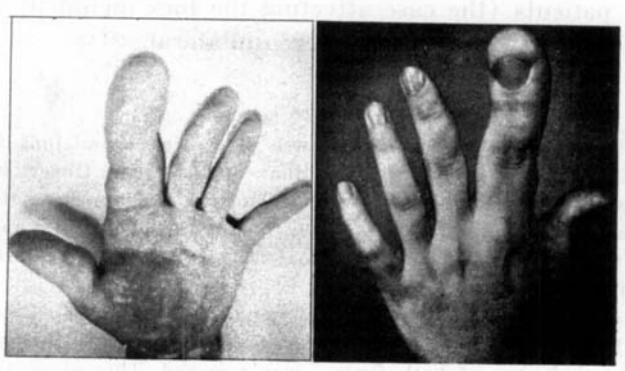
Figures 3 a and b: Case 2, female, aged 20.





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Figures 5 a and b: Case 3, female, aged 16.

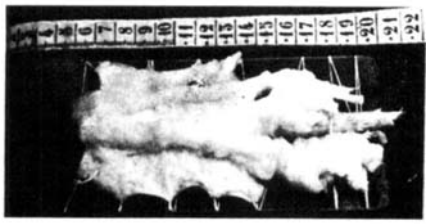
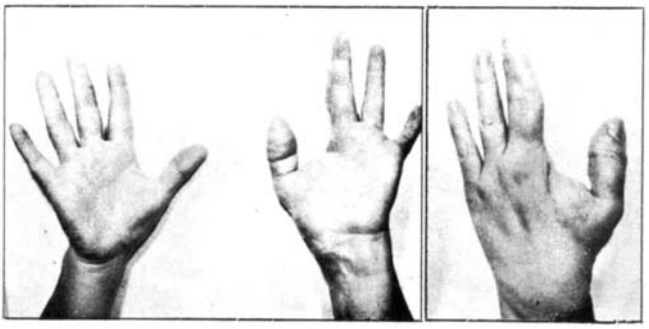


Figure 6: Case 3. Operative specimen of the enlarged median nerve with branches.



a *b*

Figures 7 a and b: Case 3, at 28 years of age.

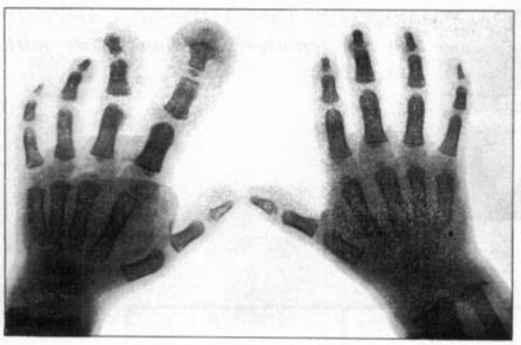
the thumb and index finger. Therefore, at 38 years of age, she had another operation, comprising resection of the hypertrophic median nerve and transmetacarpal amputation of the index finger. The median nerve was as thick as a thumb, the digital nerves were 3–4 mm thick. Micr. exam.: Lipoma.

At follow-up, 6 months after the operation (Figures 2 a and b) the main complaint, apart from the cosmetic aspect, was weakness of the thumb and deficient opposition. There had been no further growth.

Case 2.

A woman, now aged 33, who had from birth had a large right thumb which had steadily increased in size. At the age of 20 the finger showed the appearance illustrated in Figures 3 a and b. Partial excision of the increased soft tissue was done. The branches of the median nerve were as thick as lead pencils, the sensory nerves to the thenar eminence 5 mm thick. Large masses of lipoma infiltrated all muscles in the web space between the thumb and index finger, splitting the muscle fibres apart. A lump as large as a hen's egg was removed. Micr. exam.: Lipoma.

A few months later exarticulation of the distal phalanx of the thumb and thinning of the soft tissues. At follow-up still thickening of the soft tissues in the thenar eminence, but during the past 10 years there had not been further growth, and the patient declared that she was quite satisfied (Figures 4 a and b).



Figures 8 a, b, and c: Case 4, a boy aged 2 years.

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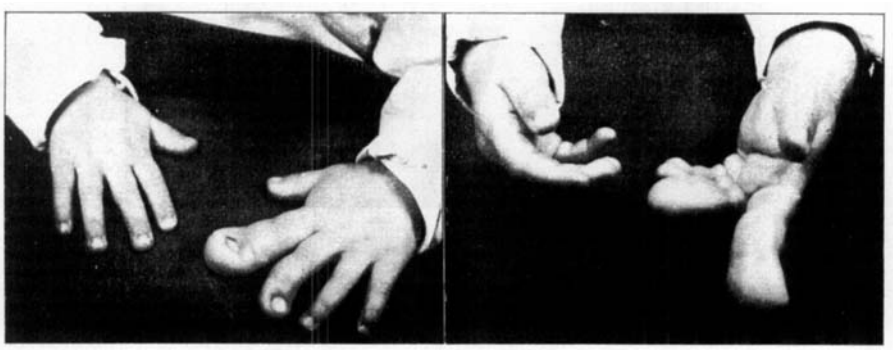


b *c*

Case 3.

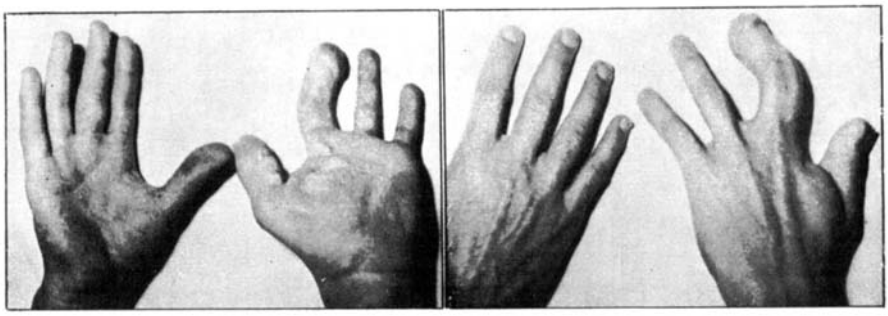
A woman, now aged 28, who had always had an enormous index finger and some enlargement of the long finger on the left hand. Referred, because of thickening of the soft tissue in the left palm and distal part of the forearm, when she was 16 years of age (Figures 5 a and b). At operation the tumour was found to consist of the median nerve which was filled with lipoma-like masses. As much as possible of the tissue was removed without injuring the branches of the nerve. Micr. exam.: Fibrolipomatous changes.

A couple of months later the index finger was amputated, and later, when she was 18, the median nerve was excised because the tumour went on growing. The nerve and its branches were found to be enormously thickened, about 3 × 1½ cm in cross section, and the digital nerves were as thick as lead pencils (Figures 6).

At follow-up now, when she is 28, she has surprisingly slight complaints on account of the lacking sensibility. The long finger had not grown further (Figures 7 a and b).

Case 4.

A male, now aged 24, who was seen for the first time at the age of 2 years. There was giantism of the left index and long fingers which showed lateral deviation



a *b*

Figures 9 a and b: Case 4, at 24 years of age.

(Figures 8 a, b, and c). Partial amputation of the index finger was done, but later at follow-up he had increasing swelling in the palm, so that at the age of 6 years he had another operation, removing a considerable fatty infiltration with widespread nerve fibres from the median nerve. However, there was still slow growth of the soft tissue, and at the age of 14 the tumour was removed together with the median nerve which was 1 cm thick at the wrist; the digital nerves were 5–6 mm thick. Micr. exam.: Lipo-fibroneuroma. Yet another operation comprised removal of a couple of neuromas as well as amputation of the index finger and of the distal phalanx of the long finger. Now, the condition is said to be stationary. The long finger which show ulnar deviation can reach the palm (Figures 9 a and b).

Case 5.

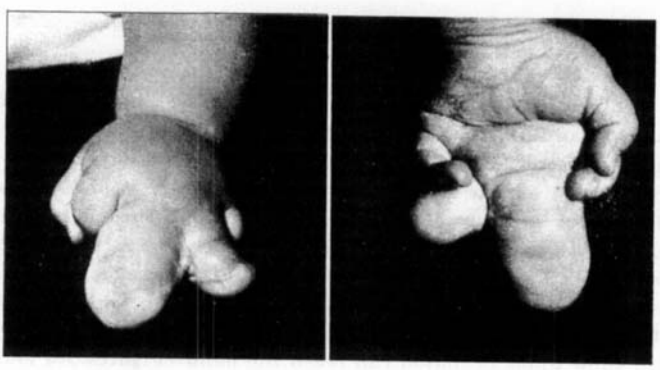
A male, now aged 21, who had from birth had giantism of the left index finger and thickening of the soft tissues of the thumb. At the age of 5 years he was subjected to operation thinning the thumb which thereafter remained fairly normal, but at 14 years of age he was referred because of enlargement of the index finger whose distal phalanx was almost spherical with 45° ulnar deviation. This phalanx was amputated, but as irregular, tender thickening remained at the tip of the stump, the middle phalanx was amputated 2 years later.

At follow-up there was no further growth of the finger, but considerable thickening of the soft parts on the volar aspect of the stump. No palpable enlargement of the median nerve.

Case 6.

A boy, who is now 7 years of age, had at birth syndactylism of the left long and ring fingers. This was repaired on the following day in the local hospital. When first seen here, at the age of 7 months, he had enormous enlargement of the long and ring fingers (Figures 10 a, b, and c). Exarticulation of the long finger was done. At this operation *no* hypertrophy of the nerve was found. Two years later the boy had developed considerable thickening of the soft tissues, and he had another operation removing lipomatous masses situated in part intraneurally in the median

a



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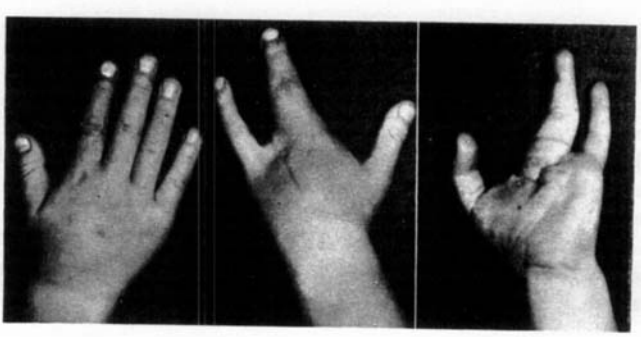

a b

Figures 10 a, b, and c: Case 6, boy, aged 7 months.

Figures 11 a and b: Case 6, at 7 years of age.

nerve. The common digital nerves to the index, long, and ring fingers were now almost 1 cm thick. Micr. exam. Connective-fatty tissue with fibrous changes.

Six months later the patient has transmetacarpal amputation of the ring finger, and at follow-up the hand was quite good. He can use it almost normally and can clench the index and little fingers completely (Figures 11 a and b).

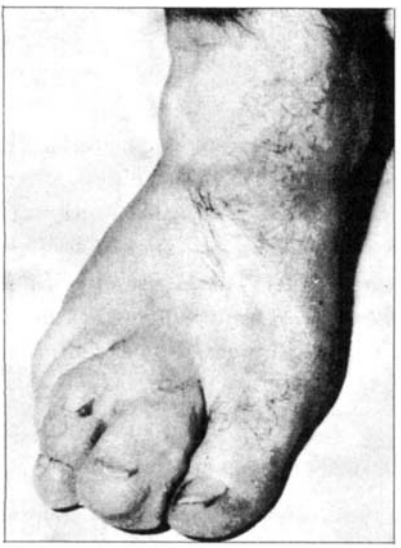

Figure 12: Case 7, a male, aged 24, with megalodactylism affecting the foot.

Case 7.

A male, aged 24, who had from birth had giantism of the right 2nd and 3rd toes, both of which are larger than the great toe (Figure 12). No definite thickening palpable along the course of the nerves. If only he has a sufficiently large shoe he has no complaints, so we did not feel that any treatment was indicated.

SUMMARY

Seven cases of the rare condition *megalodactylism* are submitted. This disease consists in local giant growth affecting one or more fingers or toes, comprising all tissues, and often associated with considerable thickening of the nerves supplying the area. The aetiology is unknown, and the histological appearances are uncharacteristic. Malignant changes have not been observed.

The treatment is restricted to plastic operation, epiphysiodesis, and amputations.

RESUME

7 cas de la maladie rare *mégalodactylie* sont présentés. Il s'agit d'une croissance gigantesque locale qui touche un ou plusieurs doigts ou orteils, qui porte sur tous les tissus et qui est souvent accompagnée d'un épaisissement considérable des nerfs dans la partie en question. L'étiologie de cette maladie est inconnue, le tableau histologique n'est pas caractéristique. Aucune modification maligne n'a été observée.

Le traitement se borne à des opérations plastiques, épiphyseodèses et amputations.

ZUSAMMENFASSUNG

7 Fälle des seltenen Leidens der *Megalodactylie* werden vorgestellt. Es dreht sich hier um ein örtliches Riesenwachstum, das einen oder mehrere Finger oder Zehen betriffts, alle Gewebe umfasst und von einer bedeutenden Verdickung der Nerven des betroffenen Gibietes begleitet wird. Die Ätiologie ist unbekannt, das histologische Bild uncharakteristisch. Maligne Veränderungen sind nicht vorgekommen.

Die Behandlung schränkt sich auf plastische Operationen, Epiphyseodesen und Amputationen ein.

ACKNOWLEDGEMENT

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