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Macrodactylism Associated with Neurofibroma of the Median Nerve*

—A Case Report—

In Hee Chung, Nam Hyun Kim and Il Yong Choi

Department of Orthopedic Surgery, Yonsei University College of Medicine, Seoul, Korea

ABSTRACT

A case of macrodactyly associated with neurofiborma of the median nerve, a congenital anomaly of the hand, affecting only one (left middle) finger is reported with a review of the literature.

Macrodactyly which is also termed local gigantism, megalodactylism, megalodactylia, or macrodactylism in other literature, is a rare congenital malformation characterized by overgrowth of one or more fingers of hand.

Macrodactyly associated with neurofibroma of the median nerve is especially rare. For this reason the following case is presented together with a review of the literature.

CASE REPORT

A 13-year-old Korean girl, was admitted to the orthopedic surgery department of Severance Hospital on Jan. 25, 1971. The chief complaints were an enlargement of the left middle finger and a soft tissue mass in the left palm since birth. Her mother stated that at the time of the child's birth, the middle finger of her left hand was much large and longer than the other fingers of the same hand. She further stated that, since the time of birth, the left middle finger had grown more rapidly.

From the age of 9, the patient developed a painless soft tissue mass in the left palm and wrist.

The past history and family history were non-contributory. The physical examination showed a moderately-developed and well-nourished Korean girl, thirteen years of age.

The middle finger of left hand(Fig. 1) shows a diffuse hypertrophy(Table 1) with normal motion, ulnar deviation of the digit, and a normal web space. There were no motor or sensory changes.

Table 1. Comparison between right and left middle fingers

	Length(Cm.)		Diameter(Cm.)	
	Rt.	Lt.	Rt.	Lt.
Metacarpal	5.7	5.7	0.7	0.7
Proximal phalanx	3.8	4.0	0.8	0.9
Middle phalanx	2.3	2.4	0.6	0.8
Distal phalanx	1.5	2.2	0.4	0.6

* This paper was presented at the 146th monthly meeting of the Korean Orthopedic Association on June 30, 1971.

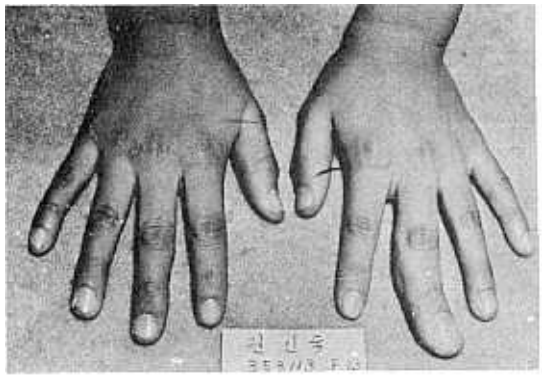

Fig. 1. Macrodactyly of the middle finger of left hand (dorsal view)

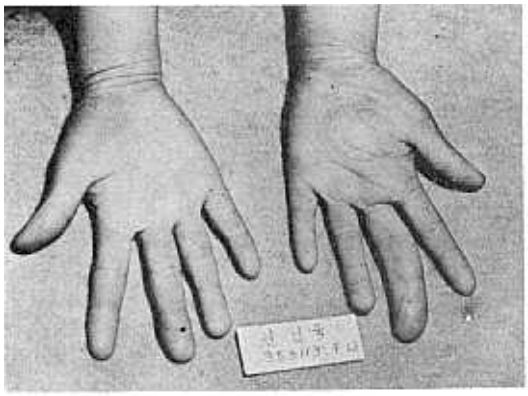

Fig. 2. Macrodactyly of the middle finger of left hand (palmar view)

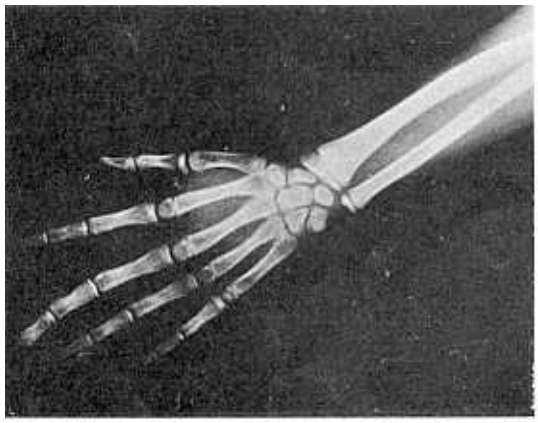

Fig. 3. Preoperative radiological findings, enlarged phalangeal bones of middle finger.

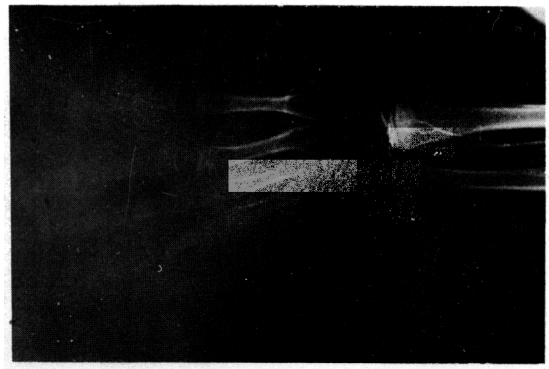

Fig. 4. Angiographic finding, scanty filling of dye distal to palmar arch.

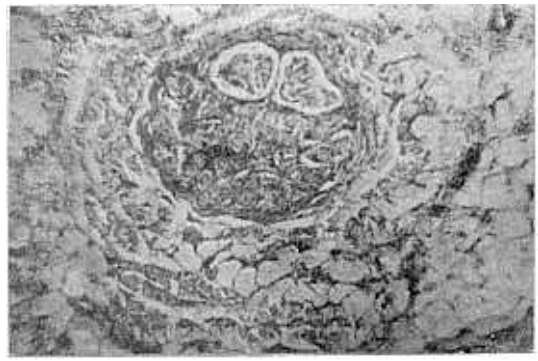

Fig. 5. Pathologic findings were proliferation of fibrous tissue within and around the nerve fibers and also increased Schwan's cells within the nerve fibers. H-E, 430×

A soft, non-tender, fixed, diffuse, and ill-defined mass was palpable in the palm and volar aspect of the left wrist(Fig. 2).

There was a Café-au-lait spot on the anterior aspect of the left ankle.

Roentgenologic examination showed the phalanges of the left middle finger to be enlarged with ulnar deviation of the middle and distal phalanges. The tuft of the distal phalanx of the left middle finger was irregular and enlarged and there was a diffusely enlarged soft tissue shadow in the left middle finger(Fig. 3).

On angiography, scanty filling of dye distal to the deep palmar arch was noticed

(Fig. 4). The routine laboratory findings were reported as being within normal limits.

On Jan. 29, 1971, under general anesthesia with a tourniquet about the arm, the entire epiphyseal plates of the distal, middle, and the proximal phalanges of the middle finger of the left hand were resected through mid-lateral incisions made on the ulnar side of PIP and MP joints and an inverted V incision at the dorsal aspect of the DIP joint of the left middle finger.

The plates were easily removed with a No. 15 knife blade. Excisional biopsy of the radial digital nerve going to left middle finger was done and the left median nerve was explored to the wrist joint.

An enlarged and hypertrophied median nerve, estimated to be 2cm. in diameter, was visualized.

The median nerve, common digital nerve, and radial digital nerve going to left middle finger showed fibromatous change. Abundant, very large, fat lobules were noticed around the radial digital nerve going to left middle finger. Atrophy of lumbrical muscles was also noticed. Postoperatively, the finger was immobilized in plaster for 2 weeks.

When the girl returned for re-examination on Aug. 10, 1971 function of the hand was normal.

Pathologic findings included proliferation of fibrous tissue within and around the nerve fibers, and increased Schwan's cells within the nerve fibers(Fig. 5).

DISCUSSION

Since the first report on macrodactyly in 1840 by Power[2] there have been other cases reported.

In 1967, Barsky[2] reviewed 64 macrodactylies collected from the last 140 years.

Among those cases, macrodactyly associated with neurofibroma was found in only a few cases. Thus macrodactyly associated with neurofibroma is one of the rarest of congenital malformations of the hand. The anomaly occurs more often in male, but our case is female.

Barsky[1] stated that multiple digit involvement is more than twice as frequent as single digit involvement and the index and middle fingers are most frequently involved.

In our case the only left middle finger was involved.

The phalanges, tendons, nerves, vessels, and skin are all enlarged, but the metacarpals are not affected. Some degree of lateral curvature of the finger may be present. In our case the left middle finger showed ulnar deviation. The etiology remains unexplained.

In 1843. Reid[2] stated that the cause of this malformation is environmental, but why an environmental factor should cause a unilateral defect, usually so selective that only a single digit is affected, is difficult to explain.

In 1930, Streeter[2] stated that this condition might be caused by germ plasm abnormality or pathology.

Bunnel[4] believed that the cause is an excessive blood and lymph supply through an arteriovenous fistula.

In 1942, Moore[2] stated that the nervous system exerts some controlling action on the process of growth, and that impaired nerves fail in this function, resulting in uncontrolled or uninhibited growth.

Friedlander[6] reported that intraneural lipoma of the median nerve was one of cause of macrodactyly.

In our case we considered that macrodactyly developed due to a neurofibroma.

There are two general forms of macrodactyly.

In one type the enlargement is present at birth, and increased size does not increase disproportionately with growth. (static type of De Laurenzi).

The second type is complicated by overgrowth of fatty tissue in the palm, the dorsum of hand, and the forearm.

We think this case belongs to the static type. The overgrowth may or may not be associated with other pathological processes such as neurofibroma, lymphoma, angioma, or arteriovenous fistula, and the skin and subcutaneous tissue are usually hypertrophied to a varying degree.

Operative treatment varies with the type, extent, and severity of the malformation. Treatment, as a rule, had been restricted to amputation, partial amputation, or non-intervention but Bunnell[4], Clifford[2], and Jones[7] suggested that growth might be checked by destroying the epiphysis.

The results of treatment in patients with macrodactyly are not entirely satisfactory. The cosmetic and functional result is poor. After surgery for angioma or neurofibroma although the appearance improved recurrence is more often seen in neurofibroma cases.

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